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ABSTRACT

Medical aspects of deafness discussed are physicians, prevention, diagnosis, and habilitation. In speeches on physicians, Louis Z. Cooper expresses need for services for deaf victims of the 1964 rubella epidemic; Jerald M. Jordan discusses the doctor-deaf patient relationship; and Hilde S. Schlesinger critically views prevention, diagnosis, and habilitation of deafness. Covered in prevention of deafness are discussions of genetic and environmental factors in deafness by John D. Rainer, types of deafness by Robert J. Ruben, and an approach to diagnosis, prevention, and treatment of hereditary deafness by Alexander C. McLeod and Anne Sweeney. Examined under diagnosis of deafness are audiological diagnosis of deafness by Carl W. Fuller and a parent's experience of medical imcompetency in diagnosing deafness in a young child. In conclusion Luther D. Robinson examines medical aspects in habilitation, Ann M. Douglas presents a nurse's viewpoint on health problems of deaf people, Lawrence Newman examines the self image of deaf persons in a society attaching stigmas to deafness, and McCay Vernon discusses the deaf community's responsibility in medical habilitation. (CB)

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Medical Aspects of Deafness

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SERVING THE DEAF • MARCH 3-5, 1971
ATLANTIC CITY, NEW JERSEY

NATIONAL FORUM NUMBER IV

Medical Aspects of Deafness

Sponsored by:

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Proceedings:
National Forum IV
Council of Organizations Serving the Deaf

March 3-5, 1971
Atlantic City, New Jersey

DOIN HICKS, Ed.D., *Editor*

Art Work by Phil Lohman

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DEDICATION

In grateful appreciation of their support of the COSD, these proceedings are dedicated to the members of the Gold Emblem Club.

Abrams, Maurice I.
Adler, Gerald and Edna
American Athletic Association of the Deaf
Arbuthnot, Helen C.
Benson, Elizabeth and Mary Alice
Bible, Lenore M.
Burstein, Gerald and Theresa
Burstein, Stephen D.
Carney, Edward C. and Ruth
Cherry, Stephen L.
Clarke, Gordon W. and Ruth
Community Speech and Hearing Center, Inc.
Cranwill, Alfred and Evelyn
Curtis, Gary A. and Betty
Duning, Mrs. Harriet
English, Millard B.
Episcopal Conference of the Deaf
Gallaudet College Alumni Association
Garretson, Mervin D. and Carol
George, James
Gross, Seymour M.
Hall, Reverend Martin J.
Henderson, Rance
International Catholic Deaf Association
Jordan, Jerald M. and Shirley
Katz, Nathan L.
Ladner, Emil S. and Mary
Lapides, Michael (deceased)
Lutheran Church-Missouri Synod
Maryland Association of the Deaf
Mayes, Thomas A. and Julia
Merrill, Edward C., Jr.
Muller, Robert J.
National Congress of Jewish Deaf
Neill, David D.
Osborne, Mrs. Thomas W.
Peikoff, David and Pauline
Pennsylvania Society for the Advancement of the Deaf
Phillips, Richard M. and Ruth
Porkorny, Daniel H. and Patricia
Registry of Interpreters for the Deaf
Rhodes, Mrs. Mary Jane
Rohe, Herbert W.
Saint Paul's Episcopal Mission for the Deaf
Sanderson, Robert G. and Mary
Sonnenstrahl, Alfred and Debbie
Steideman, Mrs. Hazel
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Sullivan, Frank B.
Van Nevel, Al
Vogt, John P.
Warshawsky, Leonard B. and Celia
Watson, David O.
Williams, Boyce R.
Wondrack, Lydia M.
Wukadinovich, Michael L. and Elodie

EDITOR'S PREFACE

DOIN HICKS, Ed.D.
Dean Pre-College Programs
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Washington, D.C.

Like its predecessors, Forum IV was highly successful in focusing attention on a major area of concern relating to the well being of deaf persons. Both the formal presentations and the discussion sessions revealed significant consensus in identifying problems and in suggesting alternatives for effecting solutions to those problems.

A recurring question throughout the Forum was, "How do we reach Physicians?" This was not a question implying criticism but one with due respect to a noble profession beset with manpower and facility needs and one facing the additional burden of keeping abreast of a rapidly growing body of medical knowledge. There is a realization that the deaf population is a minuscule segment of our society and that appropriate medical services to this group require special effort on the part of physicians, particularly in the non-medical area of communication skills. For reasons such as this, it is highly appropriate that deaf persons and persons who work with the deaf make special effort to provide all possible support and encouragement to the medical profession.

Historically the medical profession has reacted very positively to the needs of the various segments of society. There was, throughout the Forum, an expression of optimism that increasingly comprehensive medical services would be made available to deaf persons, but that the responsibility must be shared by all who have concern. Herein lies the challenge–first, to those who work with the deaf and to the deaf themselves to provide the awareness and to help the medical profession acquire the needed skills and, second, to the professionals in medicine to accept the deaf individual as one deserving added time and attention.

COSD Forum IV represents only a beginning. The readers of these proceedings are encouraged to share the responsibility of reacting to the needs expressed herein.

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WELCOME

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Northridge, California

On behalf of the Council of Organizations Serving the Deaf it gives me great pleasure to welcome you to the Fourth National Forum.

I am thankful this Forum is being held in Atlantic City instead of where I just came from–the earthquake county of Southern California and, in particular, San Fernando Valley.

In San Fernando Valley, the ground is still shaking every day. Over 200 after-shocks have been recorded. I am still shaky and no doubt the same can be said of Dr. Ray Jones, Dr. Tom Mayes, and others here from Southern California.

In fact, last night I could not get to sleep until my wife rocked the bed.

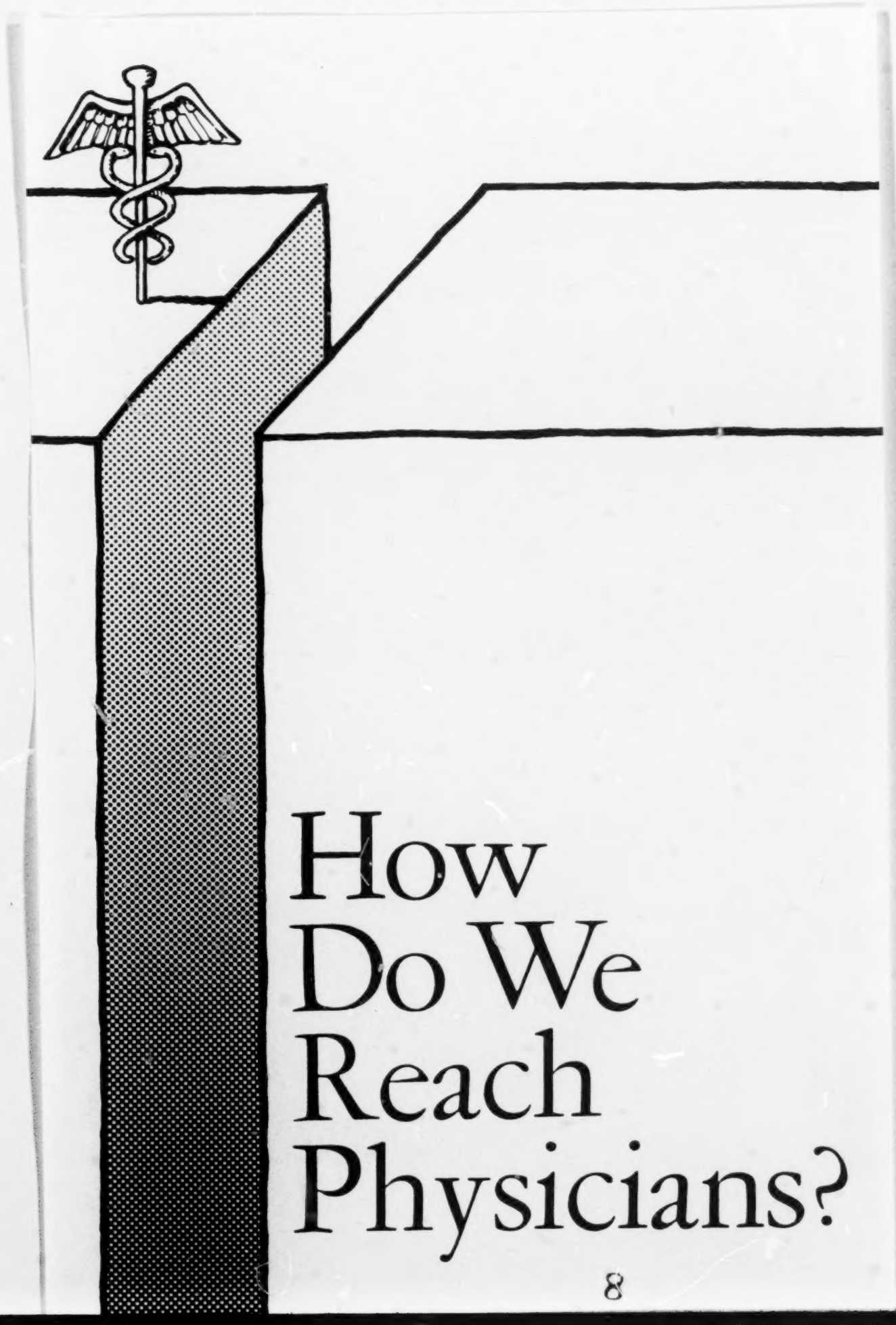
Scientists have been studying this earthquake for several weeks–attributing it to this fault or that fault. The best theory seems the earthquake is due to Agnew's fault.

Several days befory the earthquake, Vice President Agnew practiced for the Bob Hope Golf Tournament in Palm Springs. The golf "pro" kept telling him, "No, no, hit the little ball, the little ball!"

No wonder mother earth shook in protest at such treatment!

But we are not here to discuss earthquakes, no matter how interesting they may be. We are here to explore, discuss, and, hopefully, find solutions to the many problems of our theme, "Medical Aspects of Deafness." It is a challenging theme and it is our hope you will experience a profitable and enjoyable Forum.

Thank you.



How Do We Reach Physicians?

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Director, The Rubella Project
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New York, New York

DEAFNESS: ONE PHYSICIAN'S VIEW

Ladies and Gentlemen. Member Organizations of the Council. Thank you for the privilege of allowing me to open your Fourth Annual Forum devoted to the theme "Medical Aspects of Deafness." The title covers an area of great importance to all of us, and recognizing the expertise of the participants who will follow on this program, I share with you great expectations for a fruitful forum. In my own presentation, I must focus on personal experience, i.e., on the deaf infant and child, although I recognize that deafness is a problem of all ages.

When Dr. Doin Hicks called some months ago to ask me to participate, I accepted with mixed emotions. I had a great desire to share some of my recent experiences and strong feelings with you. They are relevant to the Council—but, I was pained by self-awareness that I am not an expert in any aspect of deafness!

I was reminded of an old story about Charlie, the proverbial town drunk, who came down here to Atlantic City to an annual national meeting of his "Lodge." Charlie stopped by the bar with a few friends, then decided he would go to his room for a little nap before dinner. In his ocean front room, he opened the window for a little fresh air and heard band music coming up the boardwalk. Being a lover of bands, pretty girls, and colorful uniforms, Charlie leaned a little farther out to get a better view. In his enthusiasm and somewhat intoxicated condition, he leaned too far and fell out onto the boardwalk. Fortunately, he fell only from the second floor, landed in a relaxed position on his back, and was merely stunned. A large crowd quickly gathered around the supine body and, just as Charlie roused himself onto one elbow, a policeman shoved through the crowd, looked down at him, and said, "What's going on here?" Charlie looked up, still somewhat puzzled, and answered, "I don't know, officer, I just got here myself!"

Unfortunately, I'm very much like Charlie. Until just a few years ago, like most of my medical colleagues, I was blissfully ignorant of the magnitude and the character of deafness as a problem of personal and public health. I knew a few persons who wore hearing aids, a few other

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"old timers" I wished would wear hearing aids–because I got tired of shouting–and a few peddlers who came around with cards reading, "Please buy. I am a deaf-mute." I was aware of what antibiotics had done to lessen the ravages of middle ear infection in children, that radical mastoidectomies were now rare operations, that vaccines were available to make measles and mumps rare diseases, that otologic surgeons had developed new operations to "cure" otosclerosis (whatever that was), that advances in electronics were heralded by advertisements in popular magazines for hearing aids so small they could fit unnoticed into eye glasses, and that industrial noise, jet airplanes, and rock bands could cause significant accoustical trauma.

I even included a question or two about hearing in my routine history-taking, and simple whisper, tuning fork, and wristwatch tick tests in my complete physical examinations. But I had no gut feeling for deafness, no real concern for its prevention, diagnosis, and treatment, no awareness of the habilitational, educational, and social threat which deafness posed to the lifestyle of its victims and their families.

Epidemic Altered Career

The rubella epidemic of 1964 not only altered the lifestyles of more than 30,000 families, whose new infants had birth defects from this infection acquired while still in the uterus, it altered my career as well.

At the time that epidemic was sweeping across this land, I was planning, with colleagues who had been working on rubella for a number of years, further studies to characterize the viral immunology of the disease and to develop new sera and vaccines for its prevention. The team I joined in 1964, using the new tools developed in 1962 at Walter Reed Army Hospital and Harvard University, already had completed fundamental studies in children who were allowed to "catch" rubella under conditions of close observation and isolation. These studies called into serious question the usefulness of quarantine and gamma globulin treatment for the prevention of rubella or its consequences. They demonstrated that the person with rubella had virus present in his blood (viremia) and pharyngeal secretions for as long as a week before onset of his rash and in his pharyngeal secretions for one to two weeks after the rash began to fade. In other words, the patient was contagious before he was aware he was sick.

Of great importance, if the patient were a pregnant women, the virus would be present in the bloodstream with access, through the placenta, to the developing fetus, even before she was aware of her infection. Infection, with viremia, pharyngeal shedding, contagion, and risk to the fetus would occur with or *without* the presence of a rash. In other words, the patient could be in trouble without realizing it. Of equal importance, the studies showed that a person who had rubella developed specific antibodies, detectable in his blood serum shortly after appearance of the rash, and that these antibodies, which lasted for years and in most instances for life, were protective against another episode of the disease. It was this

observation that raised hopes for an effective vaccine. Rubella behaved like certain other systemic or generalized infections known to be preventable by live attenuated virus vaccines, namely poliomyelitis and measles. An attenuated vaccine is a live virus which has been weakened in the laboratory so that it no longer causes illness, but still provokes the body to make antibodies.

You all know that hopes for an effective rubella vaccine have been realized. In fact, since licensure for general use in June 1969, more than 25 million children in the United States have been immunized. But that is another aspect of the story I'd like to leave until later.

What happened after the epidemic of rubella in the spring of 1964 that led to a new dimension for our rubella research and an awareness in our group of medical scientist-physicians of the needs of deaf children? The answer, very simply, was this: In our community—metropolitan New York—with a population of more than 12 million, thousands of pregnant women had rubella in the spring. Then, in the fall and early winter, those whose pregnancies were not terminated by therapeutic abortion came to term, delivered their babies. Although many appeared to be perfectly normal in the hospital nursery, several hundred others had obvious problems such as hepatitis and purpura, which is a result of a deficiency in blood platelets.

Since the work of the New York University Medical Center-Bellevue Hospital rubella research program was known to many doctors in the community, we were asked to see many of these infants to help in diagnosis and initial management. Among the first things we learned was that these infants were still infected with rubella virus, and still contagious to susceptible persons who came in direct contact with them. We now know that in most infants this period of contagion lasts for just a few months.

Rubella Team Assembled

With the help of emergency funds from the local chapter of the National Foundation-March of Dimes, a team of medical specialists was assembled as the Rubella Birth Defect Evaluation Project. A training grant in Pediatric Audiology and another for psychiatric-psychological evaluation were provided through the Children's Bureau and, working with the large medical community in New York, we were in business for defining and providing care for the medical needs of rubella victims. Since 1965, more than 700 children have received service, the majority on a longitudinal, ongoing basis.

High quality, coordinated, sophisticated medical, psychiatric, and audiologic services for rubella children are not easy to provide. Although a communication disorder was the most common single problem, most of our children were multiply handicapped. Heart disease, cataract, glaucoma, and encephalitis (infection and inflammation of the brain) were present in varying combinations in many of the children. The encephalitis, which

could involve any area of the brain, left a spectrum of damage ranging from profound global retardation to cerebral palsy with spastic diplegia; behavioral disturbances, mild (such as hyperactivity) to severe (such as infantile autism); perceptual problems with subtle learning disability; and complex central auditory impairment with severe receptive and expressive language disorders. With all these problems, we were grateful for children whose *only* problem was sensorineural hearing loss.

You are all aware of two important facts: 1) that the first years of life represent a unique critical period for auditory and language learning; and 2) that the rubella "deaf" child, like most of our country's three million hearing-impaired children, is uncommonly without some sensitivity to sound. We were aware, also, of these "facts of life." As soon as medical crises were stabilized, we worked hard with our colleagues in hearing and speech clinics throughout New York to refer rubella infants and their families for appropriate amplification and auditory training. New York is unusually fortunate in the number and variety of services for handicapped children. The often maligned, but often far-sighted Board of Education pioneered by placing teachers of the infant deaf in hospital-based hearing and speech clinics more than 15 years ago. The private schools for the deaf in the area also have a tradition of responsiveness to community needs.

Despite unusually cooperative collective efforts by representatives from health, education, and social service agencies, by 1967–when our "rubella 1964" victims were already three years old–more than one-half were receiving no educational service or service too insignificant to be meaningful to the child or his family. Naturally, the children in greatest need, whose handicaps were often multiple and whose total family resources were most limited (intellectually, socially, and financially), were those least likely to be receiving service.

The frustration and anguish that families experienced became our own as we listened to them, observed their children, and struggled with the professional community to expand services before irretrievable ground was lost.

As you might expect, from experience in your own communities, we did lose much–critical periods for hearing and language, emotional overlay, and family disruption.

Enormous Progress Made

It is no comfort to the losers, the now nonverbal children who had the potential for integration into the aural-oral school community, the children originally mislabeled and misserviced as retarded or emotionally disturbed, and their families, that we have learned from this experience of a single epidemic. Enormous progress has been made during the past five years both in the status of our services for special children, and in our concepts and our current expectations.

Tangible evidence of this progress is extensive. Familiar examples include: amendments to the Elementary and Secondary Education Act

authorizing a range of special educational programs, creation of the Bureau of Education for the Handicapped in the United States Office of Education, The Model Secondary School for the Deaf, the Deaf-Blind Centers Act creating regional centers for services to these multiply handicapped children, the Handicapped Children's Early Education Assistance Act, and the new Developmental Disabilities legislation which broadens the mandate of the Rehabilitation Services Administration. All of us share the responsibility to work toward "full funding" for these important ventures. As you know, it is important to see that Congress appropriates and that the Executive Branch allows expenditure of the funds authorized in the initial legislation creating these programs.

Less tangible, but no less important, are the indications of improving communication among different disciplines, as seen by such efforts as the recent *Report of the Joint Commission on Mental Health in Children.* This report took a broad view of what child health really involves and presented an imposing challenge for the coming decade. My presence on this program here today is an example of the new cooperation between disciplines.

Our President, Richard M. Nixon, has recognized the first years of a child's life as being critically important for the well being of that child and of our Nation. He has expressed clearly his belief in the value of local involvement for establishing priorities, identifying problems, and seeking solutions–a trust in our federal system of government. It is important that we help him to translate these beliefs (which I share and, I suspect, most of you do, also) into action.

One of the most useful methods for evaluating "action" is to look at where money is going: for our purposes, to the federal budget. We all recognize that a disproportionate share of our federal taxes is spent by the Department of Defense. Even the most militant among us (a small minority, I'm sure) accept this at best as a "necessary evil" of living in a difficult world during difficult times. No one questions the desirability of reducing these expenditures. It is not appropriate in this Forum to discuss the controversial details of how to do it. (Certainly, Mr. Melvin Laird, long a strong supporter of health research and services before he became Secretary of Defense, is grateful for all good suggestions on this point.)

Coordinated Programs Needed

However, there is another area of funding where the relative federal expenditures may surprise you. They did me. Despite our child-oriented society and "priority commitment to children," nine dollars per person per year is spent for the aged for every one dollar per child! I do not believe our senior citizens are blessed with a surfeit of services. In fact, for many, what is available is inadequate. But, I believe this comparison does expose a myth for what it is. Being an immature society is not the same as being "child-oriented."

What am I driving at?

All of us would like to see coordinated programs for delivery of full services to children and families. For the child with congenital hearing impairment, this means early identification, proper diagnosis, amplification, parent education, schooling, and follow through, beginning in infancy. We are far from achieving that goal. What can we do?

1. Voice our concern continuously. In our democratic society politics, defined by Harold Laswell as "Who gets what, when and how," is still the name of the game. The deaf are a small minority. To get a fair share for their needs, the deaf and those concerned for them must expend heavy effort in public and professional education in order to create and maintain an enlightened community of neighbors and voters. Educating doctors is one small but essential ingredient. An important tactic is the reliance, not only on humanistic values, but on solid cost-benefit analyses of the long-term payback to the community of adequate services compared to inadequate services.
   Reliance solely on the "vending machine concept" (as John W. Gardner has pointed out) is doomed to failure. That is the concept based upon "put in a coin, and out comes a piece of candy" or "pass a law and out comes a solution."
2. Generalize our concern to encompass a larger constituency. This is important to the deaf for two reasons. First, it provides a broader political base. Second, and more important, the problems of the deaf are unique only in a limited way. The deaf share many problems with others who have organic and/or functional impairments, and have major needs which are common to our society as a whole.
3. Be alert to the danger of simplistic solutions. The concept of a monolithic, well-organized national structure for delivery of all human services is attractive, especially to compulsive professionals (a category into which most of us fit), i.e. the often alluded to "Scandanavian system." We are all aware of the fine work done in countries such as Sweden, and we can and should learn from them. Nevertheless, the differences between an established, homogeneous country of eight million persons and a developing heterogeneous nation of 200 million persons are too obvious to be reviewed this morning.
   One simple example may help to remind us of these differences: I have been working with local, state, and national agencies, governmental and private, to expedite distribution of rubella vaccine. A reasonable sum of federal funds was diverted to this purpose and channeled through an efficient agency, the Center for Disease Control (CDC) for distribution to the individual states. Federal guidelines, obviously, were the same for all local communities. A map was prepared recently by CDC and is color-coded by state to indicate the percentage of children now vaccinated. The map shows that a wide variation exists, ranging from less than 10 percent in several states to greater than 90 percent of the target population vaccinated in several others. Distribution of rubella vaccine is simple in comparison to

provision and distribution of adequate services for deaf and normal children.

A monolith, even a benevolent one, is not politically feasible in our country and, considering the state of knowledge concerning people-oriented issues, most of us would agree that a monolith would be undesirable. There is safety and strength in our current diversity, which is reasonable compensation for definite sluggishness of response.

Who Takes Responsibility?

If not a monolith, then who will and should take responsibility for our deaf children and adults and for our other citizens with special needs? This is a serious and difficult problem, one that causes me particular discomfort as I review the histories of many patients–especially children with hearing loss and central language disorders complicated by additional impairments.

Delayed and inadequate service has been compounded at all levels. Physicians have been slow to refer or to follow up on referral to hearing and speech clinics. Hearing and speech clinics, having legitimate diagnostic difficulty, have delayed in referral to educational facilities, and the educational facilities have often been reluctant to accept these complex children. Unquestionably, facilities–especially private ones–have the power and duty to define the nature and limit of services they will deliver. I have been cheered by the willingness of certain facilities in our New York area to accept and try to work with difficult children and their families.

On the other hand, I have been distressed by the refusal to bend of other facilities and by their cavalier method of refusal: Often, "Your child is not suitable for our facility. Why don't you try the agency down the street?" All too often, no checking was done to ascertain that the agency down the street was in any better position and no follow through undertaken to be sure the bewildered family obtained service elsewhere.

At another level, responsibility has not been managed with great success. The rubella epidemic began in early 1964 along the east coast of the United States. Medical and public health authorities recognized its magnitude and consequences by mid-1965. However, in most states, it was two to five years before educational agencies began to gear up in earnest for service to its victims. The communication between health and education was, and still is, inadequate–although it is getting better. This gap is especially disastrous in the field of hearing and language-impaired children.

Since an "umbrella agency" for human services is out of the question (in fact, it would have to be broken down into sub-units to function), what can be done? To me, the most attractive approach is one of "centers of responsibility" and of "shifting leadership." The sick infant or child obviously requires medical treatment, with a physician establishing priority. However, as the health need is brought under control, social, environmental, and educational needs become predominant. There should

be smooth transitions from one leadership role to the next. Equally obvious is the constant overlap which should be part of comprehensive service. This requires a degree of cooperation that is still honored primarily by lip service in most areas. I have reviewed health records, checked on evaluation of hearing aids, eye glasses, immunizations, and social service support in numerous educational facilities and too often have been disappointed. Having a school nurse and school doctor or panel of specialists is no guarantee of quality service—just as a Department of Otolaryngology was no guarantee when I was a student in medical school that I would learn anything about deafness.

Survival Skills Raised

As life has become more complex, survival skills in society have been raised constantly to more sophisticated levels. When all is well and goes well with a child and his family, they both can acquire the necessary skills to keep up. But the balance is now tenuous; the margin of safety is narrow. Physical impairment or social, family, or environmental mishaps occur, and many Americans easily fall by the wayside. These disasters can be prevented only by adequate compensatory help, available *when* it is needed.

Those of you here have years of collective experience and feeling for providing a compensatory service to persons upon whom fate has intruded. You know from personal experience the strengths and weaknesses in your own communities, just as I do. I believe this awareness presents a responsibility to build now on what we have. This will require a sharing of knowledge and a social maturity which has been lacking to date. Vested interests, guilds, false professionalism, generalization of expertise in one field in an attempt to dominate at the interfaces with other disciplines, and failure to remember that the only "reason for being" of any provider of service is the "need of a consumer" are issues with which we must come to grips in this decade.

No single individual, discipline, or community has the competence to provide full service. To illustrate the complexity and dangers, I should like to cite one last example of how our own program has failed. This mother, as a teenager, delivered a brain-injured, hearing- and vision-impaired child as a consequence of rubella in 1964. She lives in an urban black ghetto, on welfare, getting clinic-type care. Her second son came two years later, and then a daughter—without a stable father-figure in the household. So, here is a mother overloaded with responsibility. Her rubella son will drift off to institutional care. Her second son, once a beautiful child, is now severely retarded from lead poisoning. And I am wondering what will happen to the baby. Proper service, properly timed, could have averted this familiar tragedy. We are not even close to providing such care for our well-endowed citizens, let alone the poor.

Because of your personal experiences, skills, and commitment, I look to each of you for leadership. Until we can provide for these deaf children,

poor children, and others on the margin, we cannot fulfill our own potential as individuals and as a nation.

Thank you for allowing me to share with you what one doctor has learned from deafness.

ACKNOWLEDGEMENTS

The views presented in this paper are my own. However, the substance of my presentation was provided by many patients, families, and colleagues who have shared their experiences and knowledge with great generosity. Our rubella studies have been supported in part by funds from the National Institute of Allergy and Infectious Diseases (Grant AI-07578 and Contract 69-2222 from the Infectious Disease Branch), the National Foundation, and the Health Research Council of New York City (Career Scientist Award I-526).

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"DOC, I CAN'T HEAR SO GOOD"
A Deaf Consumer Views the Medical Profession

A great deal has been written about early diagnosis of deafness. Much is being done in the field of prevention. Workshops have been held on the subject of mental health for the deaf. Yet Nanette Fabray, the noted entertainer who has a hearing impairment, in an address to the AMA Congress in Los Angeles last fall, said, "Too many doctors know too little about the nature of hearing disorders, the psychiatric trauma involved, the life experience of those affected, or even the extent of remedial techniques available." She then went on to give a very fine presentation of the educational problems involved.

I would like to devote most of my time today to a much neglected area of medicine, that of the doctor-deaf patient relationship. Like most persons, the deaf individual tends to look upon his doctor as being something just a step below God. Again, like most people, the deaf person is not inclined to question what his doctor says or does. However, unlike most persons, the deaf patient rarely enjoys a genuine rapport with his doctor. My own doctor claims that 90 percent of a patient's problems are psychological and only 10 percent are of a medical nature. Even if you don't agree with his percentages, I am sure you will all agree that treating the whole patient is of paramount importance. The doctor with a deaf patient, however, tends to treat only the ailment.

I believe each deaf person present could regale us with stories of his problems with his doctor. There is that inevitable time when you get a chest x-ray. The technician says, "Take a deep breath and hold it." Then he disappears for ages while you slowly turn purple waiting for him to come back so you'll know it is over. And how about the eye examination—how can you possible lip-read with those drops in your eyes? Or the proctoscopic examination . . . how, in heaven's name, is the deaf patient expected to communicate in the dark, in THAT position?

Take my own case. I am what is generally known as a good lip-reader. My doctor, like so many others, is of the opinion that since I speak clearly, I must lip-read equally well. I assure you that his diagnosis is wrong. He hates to write, and I strongly suspect that all those stories about doctors' handwriting are true. Many a time I have struggled to lip-read him, pretended vast understanding, and then gone home to have my wife call his nurse to find out what the diagnosis was.

Lack of Rapport

The main reasons for this lack of rapport between the doctor and his deaf patient appear to be the non-orientation of the medical profession to deafness and its non-medical complications, and the ever-present communication problem.

I once asked a young doctor just what he had learned about deafness during his training. He said he could recall learning quite a bit about the causes and diagnosis of hearing loss and the anatomy of the ear. But as far as he could remember, nothing was ever said about the possible problems inherent in the doctor-patient relationship. Unless he happened to encounter a deaf patient during internship, the aspiring doctor would never have the slightest inkling a problem might occur in communication.

I would like to present two hypothetical cases: You, as a doctor or pediatrician or otologist or whatever you are, have a young patient with a newly-diagnosed case of irreversible hearing loss. Now what?

You can, and should, refer the parents and patient to the nearest speech and hearing clinic. There they may get the proper information on the feasibility of hearing aids, on the value of speech training, and on the available educational resources. But that is only part of their need. They are going to look to you for reassurance and continuing guidance. After all, you are the one they have depended upon in the past. Further, you will most likely have the medical responsibility for that young deaf patient for many years to come. Are you aware of and prepared for the many emotional and psychological problems that may be involved?

Since in most cases it is the doctor who has the first professional contact with a hearing problem, I believe that he must be given the necessary information to properly advise the patient and/or parents. It is not sufficient to refer them to an otologist or an audiologist and let it go at that. The doctor, ideally, should be well informed about hearing aids, speech training, and educational methods. But that's probably asking too much of the average doctor. He should, however, be much better informed than he is now. He should be aware of the nature of the Council of Organizations Serving the Deaf and be able to refer the client there for further assistance and information.

The other hypothetical case: A deaf adult enters your office seeking treatment. What do you know about the problems of communicating with him? Please don't tell me that *all* deaf persons can lip-read (one doctor actually said that!). In many instances, even communicating by writing may not be as easy as you might expect. I would like at this point to stress that deaf persons, like non-deaf persons, are highly individualistic. It is wrong and futile to try to cram them into a single mold. Generalizations are always dangerous. Nevertheless, let's face it—when a person loses his hearing at an early age (say, before five) it is practically inevitable that he will have some degree of language deficiency. Since most profound deafness occurs at birth or shortly thereafter, the chances are that your patient will be one who has difficulty in understanding or expressing

himself in the English language. Please do NOT confuse this difficulty with lack of intelligence!

First Encounter Frustrating

Depending on your patient's degree of language deficiency, your first encounter is going to range from frustrating misunderstanding to utter confusion. Terms which you routinely use most likely will be completely foreign to him. A deaf friend of mine once asked me what it meant when the doctor asked him if his pain was sharp or dull. He had no idea what the two words meant in that context. Lord knows what answer he had given or if the doctor had been misled by it. Just think what happens when you use such words or phrases as "pulmonary infection," "edema," "stool," "carcinoma," or "congestion," to name just a few.

Then, too, your patient may use some rather down-to-earth words in describing his problem. This may lead you to form an entirely wrong concept of his intellect, character, and socio-economic standing. The sad fact is that those will be the only words he knows and he will be unaware that they are improper. Since no one has ever taught him the more socially acceptable words, your use of such terminology will only compound his confusion.

The best way to communicate with such a patient is through the language of signs. It is hardly realistic to expect all doctors to learn the sign language. It appears reasonable, however, to expect them to be aware of the problems they may face and alerted to the fact that interpreters can be of great help in such cases.

Doctor-patient rapport is greatest where communication is free and easy. Nowhere is this more vital than in the field of psychiatry. Unfortunately there are very few psychiatrists who can communicate through sign language. Where can the language-handicapped deaf person obtain psychiatric help? Unless he lives in the metropolitan areas of New York, Chicago, or Washington, or the few other places that have developed programs to help the deaf patient, he is out of luck. Indeed, there are many documented cases of deaf persons being placed in institutions for the mentally ill when their only real problem was their inability to express themselves. And who had a part in placing them there? A doctor who had misunderstood their language handicap (and emotional disturbance) for mental illness.

What happens to the deaf patient in an emergency ward? There is no time for misunderstanding yet it isn't possible to lip-read a person wearing a surgical mask—and a pad and pencil are hardly sterile.

Improving the Situation

I have spoken about some of the problems involved in the doctor-deaf patient relationship. Now I would like to present some suggestions for improving the situation.

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First and foremost, I suggest a series of pamphlets produced by joint COSD-AMA effort orienting the medical profession to the problems. The pamphlets should provide data on where further information and assistance can be obtained. They should be distributed to all practicing doctors' offices and routinely distributed as part of medical school literature.

Second, the medical profession needs to be informed of the availability and desirability of interpreters in those cases where communication is vital. There should be no stigma attached to using an interpreter nor should it be felt that such use infringes upon the privacy of the doctor-patient relationship.

Third, the profession needs to do much more for deaf persons in the field of mental health. It needs to train personnel specifically for this type of work. This will involve, in addition to the customary psychiatric training, intensive training in the language of signs. Early decision by medical students to work with the deaf alone or with the general public as well will facilitate sign language acquisition and, to this end, suitable publicity must be given to the trainees.

Fourth, the AMA and COSD should collaborate on a "When you see the doctor" pamphlet for the language-handicapped deaf patient. This could include, in simple terms, descriptions of the more common ailments and perhaps illustrations of the pertinent areas and procedures.

Publicity in the AMA Journal would also be of great help in bringing all this to the attention of the profession.

Finally, here are suggestions that a doctor can implement on his own without waiting:

Instruct your receptionists and aides to explain to the deaf patient the reasons for unexpectedly long waits beyond appointment times. It is a small courtesy but serves to ease the tedium of the wait and assures him that he is not forgotten.

A short written explanation of a procedure will serve to relax a deaf person's tension—it's the not knowing what it is all about that is nerve-wracking.

Keep written matter in layman's terms, please. Clarity is more important than anything else.

Don't assume that your language-handicapped patient knows what you are saying just because he nods his head. Ask questions to satisfy yourself that he comprehends.

Explain referrals clearly and give reasons. Otherwise they are apt to cause concern.

Prescriptions should be explained and possible side effects delineated.

If a deaf parent brings in a hearing child, be sure to address yourself to the parent and not to the child even if the child is interpreting for you.

If you are uncomfortable with a deaf patient, try not to show it. He has enough problems without thinking that his doctor doesn't like him.

Keep in mind at all times that communication means the exchange of thoughts and if you aren't getting through, regardless of the method, you aren't communicating!

Above all, remember this: a smile works wonders and is understood by anyone. Facial expressions convey more to a deaf person than anything you do or say. So if you happen to be thinking about that missed putt on the seventh hole, smile anyhow!

A Friend's Experience

I think what I have been saying can best be illustrated by the experience of a deaf friend. This man became deaf as a result of illness in his teens. He has excellent speech and is also a good lip-reader. He had established a fine rapport with his doctor. One day his wife, who is not so fortunate and can neither speak nor lip-read, visited the same doctor for the first time. That evening she told her husband that she could not understand why he liked the doctor so much. Her own impression of him had been decidedly negative. He had frowned a great deal and had appeared angry that she could not understand him as well as her husband could. She had left the office with a prescription but without knowing the diagnosis and without any directions other than those on the prescription. Her husband, quite upset about this, went to the doctor's office the next day. Since the doctor was out, he wrote a long note explaining his and his wife's feelings about her visit.

That same evening the doctor came to this man's home and apologized. He had not realized how his actions had appeared to the patient. He had been a bit nonplussed by the woman's inability to understand him with the same ease as her husband, and had taken refuge in brusqueness. After a friendly and informative discussion, the doctor gained a true insight to the situation and both my friend and his wife now enjoy the same warm rapport that should permeate all doctor-patient relationships.

All deaf persons haven't been this fortunate. There is no reason why they shouldn't be.

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PREVENTION, DIAGNOSIS, AND HABILITATION OF DEAFNESS: A CRITICAL LOOK

"I am bound by my own definition of criticism: A disinterested endeavor to learn and propagate the best that is known and thought in the world."

Matthew Arnold

Essays in Criticism: First Series 1

I dedicate this Chapter to medical students–who will soon be of help to anxious parents of hearing-impaired youngsters, and who frequently are expected to be authorities without adequate training or exposure to either the diagnosis or prognosis of early childhood deafness. (The discomfort produced by the discrepancy of expected expertise with accompanying competency is well-known to the author.) In this light this Chapter will be more concerned with those areas which may be of immediate interest and use: i.e., the prevention of ototoxic deafness receives a disproportionate share because of its preventive importance, whereas the actual diagnostic procedures are only touched in passing, since they are not and need not be in the general physician's area of expertise.

Introduction: If we accept the above definition of criticism this forum can be seen as the springboard for the best that is known about the *prevention* of deafness, particularly the prevention of the handicapping concomitants of deafness caused by potentially remediable conditions. This meeting also can contribute to facilitating early *diagnosis* of deafness by pointing to those aspects of diagnosis which appear to have been little known or understood previously. Most of all, however, this yearly forum of COSD can act as a platform to "propagate the best that is known and thought in the world" about the *habilitation* of deafness.

Definition: Deafness can be defined as a stress producing hearing loss from birth or early childhood rendering a person incapable of effecting a meaningful and substantial auditory contact with the environment (Rainer, et al, 1963 p. XIV). It is also a cultural phenomenon in which social, emotional, linguistic, and intellectual patterns and problems are inextricably bound together (Schlesinger 1969). Although deafness does of course occur throughout the age span of man and is stressful at any time, the complexities of pre-lingual deafness are the most intricate and the least well understood.

Prevention

It is generally assumed that prevention of deafness is a desirable medical goal as would be the prevention of any illness. However, deafness has cultural concomitants which can partially be expressed by the following: Several deaf parents have openly expressed a preference for deaf offspring. A deaf participant at this meeting has asked the poignant question: "Should we really prevent deafness? There may be a valid reason for our existence?" However, this participant was the son of deaf parents and may have experienced his developmental stages more optimally than is the case for the majority of deaf children.[1] In spite of some possible qualifications it is usually assumed that the prevention of the chasm which exists between deaf potential and deaf achievement is a desirable medical and human goal.

How Have We Fared with Prevention: It is difficult to evaluate the success or failure of the prevention of deafness since (1) precise information on prevalence is unavailable; (2) definitions of deafness may not be the same in different studies; (3) no census of the deaf population has been published for many years; (4) the causes of deafness have changed with advances in medical knowledge. Generally it seems that, contrary to general impressions, most surveys do not indicate that the incidence of deafness is either increasing or decreasing. Perhaps advancements in treatment prevent deafness as often as they result in survival with deafness. (Myklebust, 1960 p. 30.)

Early childhood deafness can be subdivided into three broad categories each with accompanying preventive thrusts: deafness occurring prenatally (presently the largest group), at birth, and postnatally. An additional category crossing all of these is that of unknown etiology.

Unknown Etiology: Specific etiologies will be discussed under special subheadings. Suffice it to say that the incidence of etiologies is subject to as much variability as the overall prevalence. Many authors report that 30 to 40 percent of deafness is of unknown etiology, (Myklebust, 1960 p. 30; Vernon, 1969 p. 43; and Hicks, 1970 p. 86). Brown[2] (1967) assigns a smaller role (18 percent) to unknown causes and a larger role to genetic reasons. On the other hand, Meadow (1968), states

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[1]See Schlesinger and Meadow, 1971 pp. 5-23, for a discussion of the impact of deafness on the developmental process.

[2]Brown, 1967 p. 178, has an elaborate table of prevalence of deafness per 100,000 of population covering foreign countries from 1940 to 1964 and three United States Surveys. The prevalence reported varied from 39 to 160 per 100,000. Considering the usual estimate of the number of deaf individuals within the United States (c. 200,000) an incidence of 80 per 100,000 (reported by Rainer, et al, 1963, p. 66) seems reasonable.

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that more than 80 percent of the parents interviewed in her study had no definite information regarding the cause of their child's deafness. Thus, apparently a large number of cases with unknown etiologies is of specific medical importance since many of the conditions might be remediable if understood. Furthermore, etiological uncertainty has been shown to be associated with greater parental adjustment problems (Zuk, 1962; Meadow, 1968; Schlesinger and Meadow, 1971).

Etiology and Prevention of Prenatal Deafness: The causes of congenital deafness may be grouped under five major sub-headings: heredity, prematurity, maternal rubella, RH incompatability, and ototoxic drug use during pregnancy.

Genetic Bases of Deafness: One of the speakers at this Forum, Dr. Rainer, is an expert in the area of genetics and was associated with the late Dr. Kallman in some seminal work on the genetics of deafness. He will undoubtedly clarify some of the intricate aspects of this intriguing specialty. We have encountered both blissful ignorance and haunting fears on the part of parents-to-be which have clearly pointed to the need for expert genetic counselling which frequently is not available.

Numerous surveys about hereditary deafness, Rainer, et al, (1963), Kloepfer (1970), and Brown (1967) tend to indicate that from 46 percent to 60 percent of all cases of severe hearing loss are genetically determined with 73 genes apparently implicated (Kloepfer, 1970, p. 18).

Brown indicates that in all childhood deafness 15 percent is produced by several simple autosomal dominant genes with average penetrance of 80 to 100 percent; 40 percent of the childhood deaf result from homozygosity for a simple autosomal recessive gene of which there are estimated to be at least 30; while less than two percent are the result of sex-linked or sex-influenced genes.

Most hereditary childhood deafness is not associated with an identified syndrome. Thyroid metabolism defect with or without goiter is the most commonly associated trait and appears to result from a variety of thyroid metabolism deficiencies of the recessive type. Disturbances of pigment metabolism are the only syndromes yet associated with dominant childhood deafness, (Brown, 1967, p. 200).

Vernon (1969) has carefully studied a sample of 1,468 deaf students who attended or were tested at the California School for the Deaf, Riverside, during the years 1953-1964. Unless otherwise stated the incidence figures for prematurity, RH factor, and rubella and ototoxity refer to his survey (pp. 42-99). Two incidence figures are assigned to each condition: the first refers to the actual incidence in the sample, whereas the second refers to the possible range of prevalence when additional factors, such as multifactor etiologies, are taken into consideration.

Prematurity: Prematurity was directly implicated in 11.9 percent and potentially implicated in 17.4 percent of Vernon's cases. This has double importance. First, prematurity is seen by Vernon (1969) to be fre-

quently associated with brain damage, aphasia, low academic achievement, and increased incidence of emotional disturbance. Second, prematurity is a potentially preventable condition. The Joint Commission on Mental Health for Children (1970) comments frequently that inadequate prenatal services exist throughout the United States, that they are particularly under-utilized by the poor, and that this absence of services is highly correlated with the incidence of infant mortality, prematurity birth defects, mental retardation, and other disabilities (p. 32).

Rh Factor: The Rh factor was directly implicated in 2.7 percent and potentially implicated in 3.7 percent of the sample. Although the incidence is small it is of disproportionate importance if other data from Vernon is examined. He states that this group represents a high incidence of multiple handicaps (71 percent), of cerebral palsy (50 percent), and of organic involvement as found on psychological testing. Of interest is the further notation that "these children were surprisingly free of extreme emotional disturbance." (p. 110).

Since prenatal testing for Rh factor exists throughout the United States, care must be taken that all pregnant women receive adequate prenatal care. Furthermore, according to Goodhill (1967), it must be noted that other blood incompatibilities may account for erythroblastosis fetalis and its neurologic sequelae. Goodhill also stresses that it is not the icterus alone which can produce the difficulty, but cerebral hypoxia and anoxia can similarly be held responsible.

Preventively it is felt that there are startling advances for safe deliveries of Rh negative mothers with an Rh positive fetus. "Delivery timing, indications for fetal transfusions, and exchange transfusions are based to a large extent on biochemical information . . . the development of an anti-Rh gamma globulin which could be given prophylactically" was seen as imminent in 1967 (Goodhill) and has been used successfully since then (Friedman, 1969).

Rubella: Because of its cyclic occurrence, its frequent association with multiple handicaps and its possible eradication through immunization, rubella has been of general and practical interest in the 1970's. Vernon found the overall incidence from 4.0 to 9.5 percent in his sample. Calvert (1969) estimates:

> ". . . that there are 20,000-30,000 additional handicapped children in this country as a result of the rubella epidemic which occurred during 1963, 1964 and 1965. Most of these children will have sensory impairments . . . . 12,000 with significant hearing loss . . . . 5,500 with severe visual impairment . . . . another 1,250 with combined vision and hearing impairment, and 1,250 children with severe mental retardation and/or neurologimuscular disorders. There may be an additional 10,000 children with mild to moderate handicaps . . . . At the present cost of educating handicapped children, we estimate that costs for 13 years of basic

special education for these children will be more than $1 billion. If the needed special education is not provided, many of these children will have to be institutionalized for life; the cost of such care for only one-half of these children would be nearly $3 billion." (p. 2-3)

In view of the above horrendous figures, both in terms of human tragedy and cost, special speed must be attached to whatever Rubella vaccination programs are seen as most effective and profitable. Care must also be taken to clarify such issues as reinfection of vaccinated and naturally immune persons exposed in an epidemic (Horstman, et al, 1970). This research indicates that vaccines were readily reinfected a few months after "successful vaccination" and points the way to further necessary research in rubella vaccinations.

Careful attention must also be paid to research findings by Bordley, et al, (1967) which indicate that live virus has been found in children at birth and as long as four to six months postdelivery, that these youngsters may be suffering from a progressive hearing loss during the first two years of life, and that the greatest injury (in terms of multiple handicaps) was seen in those youngsters from whom live virus could be cultured after birth.

Ototoxic Deafness: A small, but important, number of cases of deafness are produced through the use of ototoxic drugs. Hawkins (1967, p. 156) indicates that the phenomenon of ototoxic deafness must be at least as old as the use of wormseed and Peruvian bark in the pre-Columbian pharmacopeia of the Americas. Quinine was generally known to cause deafness in 1884 and was found in two cases (.1 percent) by Vernon in 1969 (p. 45). Quinine and salicylate deafness is frequently reversible. But, as Hawkins (1967, p. 156-157) points out:

"It has remained for this generation to develop, by a sort of negative serendipity, therapeutic agents of highly specific, permanent ototoxic effect. There are the basic antibiotics, which began to appear some 20 years ago with the advent of streptomycin, which now constitute a small anti-social family, of which the most objectionable junior members–at least from the point of view of hearing conservation–are neomycin and kanamycin." (p. 10)

Hawkins goes on to say that these antibiotics should be employed with the "greatest circumspection, and only in cases where no other anti-microbial agent will serve" (p. 165). It must be stressed that all of the above antibiotics have been implicated in hearing loss in adults *and* in children born to mothers who had received these drugs.

Deafness at Birth and Perinatal Prevention: Deafness occurring at the time of birth is most often caused by perinatal anoxia and traumatic instrumentation. Although definite figures are not available for such etiologies, it seems relatively clear that the recent advances in obstetrics and pediatrics have drastically reduced both of these causative factors.

Deafness Post-birth and Post-natal Prevention: Deafness occurring after birth, due to inflammatory diseases and their sequelae, has shown the most drastic reduction of all causative factors. Many of the viral diseases previously implicated such as mumps, measles, poliomyelitis, have been eliminated through immunizations. In others, such as influenza, common cold, and chickenpox, research must still point the way. Many of the childhood bacterial diseases previously implicated have been eradicated, or their sequelae lessened through antibiotics.

Diagnosis

Time Factors: The importance of early diagnosis of deafness cannot be overestimated. Studies of cognitive development studies indicate the importance of early sensory stimulation (Hunt, 1964); Caldwell, 1967). Early diagnosis permits early habilitative measures to maximize both the auditory and the visual input of the deaf infant. Unfortunately, delayed diagnosis has been the rule rather than the exception until very recently. However, several investigators have been involved with the development of hearing tests during the neonatal period. This has generally followed two lines:

> . . . one is represented by the meticulous kinds of clinical observations being made by Eisenberg et al, (1964) and by Murphy (1962) by means of which subtle behavioral responses to various kinds of sounds are noted in a structured environment. The other line consists of gross screening procedures applicable to large numbers of infants in non-structured environments. The latter type of test has been reported by Wedenberg, (1963), Parr (1962), and Downs and Sterritt (1964) . . . . (Downs, 1967, p. 25-26.)

Hopes were high in 1967 that neonatal screening programs were rapidly becoming a feasible and effective reality. However, questions and difficulties began to emerge as the program spread. Glorig (1971) in a thoughtful summary of the status of neonatal screening procedures describes it in terms of an ongoing dilemma:

> "There is incontrovertible evidence that detection of impaired hearing in the infant prior to six months of age is essential . . . the newborn nursery offers the best opportunity . . . and once the child leaves the hospital the opportunity for routine screening is lost."

However, infant liability, false positives and false negatives, delayed onset of deafness caused by certain disorders in the neonatal period continue to plague investigators, and have contributed to the need and the formation of a Joint Committee of Neonate Hearing Screening– whose recommendations urge increased research efforts with extended follow-up studies. In the meantime, Glorig urges the establishment of a high risk register with the training of well-baby clinics in supervised screening techniques.

Procedures: Diagnostic procedures to detect hearing loss in young children is a complex process. It cannot be achieved by ticking watches,

tuning forks or handclapping behind the child. The procedures are sophisticated and include conditioning to sound for pure tone audiometry, play audiometry, galvanic skin response, and auditory evoked response. None of the highly specialized technical procedures are performed in the general physician's office. Furthermore, most investigators feel that the technical procedures do not obviate the need for clinical acumen and experience. A useful and concise description of techniques can be found in *Deafness in Childhood*, Chapter 2.

I have attempted to briefly outline the "known" medical facts about the prevention and diagnosis of early deafness. It is clear that vast medical and audiological advances have been made in the recent past. It might be postulated that with the existing body of knowledge diagnosis of deafness can be made early and conveyed to parents in such a way as to enable them to face the crisis as effectively as possible in order to proceed to the steps of habilitation.

The facts remain, however, that a relatively large number of parents encounter some difficulties in their initial contacts with professionals. The first of these may be summarized by a lack of knowledge about early deafness among physicians in general. Many medical schools do not include early childhood deafness in their curriculum: such deafness of rare occurrence is invisible and does mimic a number of other childhood disorders. Therefore, its diagnosis is arduous in the hands of those whose index of suspicion has not been raised by exposure to the condition. Results such as these are not uncommon: one-third of the parents of a group of known deaf children indicate that the first doctor consulted said that their child was not deaf (Meadow, 1968; Schlesinger, 1968). Clinical cases continue to appear in numerically small, but humanly tragically high numbers where misdiagnosed youngsters are discovered in state hospitals for the retarded.

Fellendorf (1970), in a questionnaire study of 260 hearing-impaired children known to the A.G. Bell Volta Bureau, indicates in general that the methods of detection used are reasonably prompt:

Suspected loss:

Before one year	50%
Between one and two years	40%
After two years	10%

There was no detectable trend that the age of first suspicion of hearing loss has dropped from 1961 to 1968 among the sample in this survey. (p. 10)

Fellendorf also points to the interesting fact that the following individuals first suspect the youngsters' deafness:

Parents	70%
Grandparents/relatives	16%
Doctor	7%
Friend	5%
Teacher/school	2%

This table is not only of great interest, but may present potentially immediate practical application. Our clinical and research findings corroborate the above order of general first suspecters.

Since it seems unlikely that either overcrowded medical school curricula or overtaxed medical students' tolerance levels can encompass any additions and therefore a sophisticated knowledge of early childhood deafness, it might be most effective to limit the dissemination of certain basic facts:

1. Eighty-six percent of relatives of deaf children are the first to correctly suspect deafness–ergo–if a parent suspects deafness please refer the youngster to a qualified speech and hearing center which does have the sophisticated expertise *and* hardware (neither one alone of these is sufficient) for early correct diagnosis.

2. Early childhood deafness is rarely total, and so the child may react to "the sound of a thumb run across a balloon," (Meadow 1968), or to a tuning fork, or to the sound of any other esoteric object in your office and still remain without adequate (unaided) hearing for the sounds of human speech, and be, therefore, incapable of learning speech without habilitative measures which are available in speech and hearing centers.

3. Early diagnosis of hearing loss is critical so that the above habilitative measures can be introduced at optimal times, i.e., deaf children do not outgrow deafness, nor is it true that nothing can be done for them before age five, nor must they "be sent away," all quotes by professionals as cited by Fellendorf (1969).[3]

The difficulties encountered by parents of deaf children in their contacts with physicians are not solely in the area of paucity of knowledge about deafness. Fellendorf (1969) feels from his findings that, "If an indictment of the medical profession is in order as a result of this survey, it is in the area of the handling of the parent, not the child." Parents frequently complained of lack of time, of apparent disinterest, lack of sympathy (Meadow, 1968).

These complaints have multiple sources rooted in intra and interpersonal feelings. The optimal equilibrium between professional mind and human response undergoes many changes from medical student to physician (Blum and Rosenberg). One of the less optimal resolutions of this ongoing conflict is the acceptance of a facade of neutrality, disinterest, a carefully calculated harriedness which permits "easy" escape from worried parents. Furthermore, the road towards physicianship is paved with certain ubiquitious events: an intricate thought

[3]On rereading the above basic facts which inclusion I would recommend to the curriculum committee of medical schools, I am caught in a dilemma. Their inclusion would indubitably be important, effective, money-saving, trauma-saving, etc.; nevertheless, they sound somewhat supercilious and prissy. Let me hasten to add that we have done such pilot teaching of medical students and have not been tarred and feathered.

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process in paraphrase: "I am insecure, I'll play-act secure, the patients want me all-knowing, I can't be all knowing, I'll play-act all knowing," can lead to ongoing difficulties in admitting mistakes or ignorance. Thus, many physicians may find it difficult to accept the correct parental diagnosis. A further, almost universal, human trait might be said to be a reluctance to give out bad news. No physician is spared that necessity, but the dread remains universal. Spock has been quoted innumerable times in this regard and Thomas Mann has sensitively described this phenomenon in *The Magic Mountain.*

However, it is easier for humans to absorb non-conflicted facts about the diagnosis of deafness than it is to change the behavior which is provoked by fear, reluctance, insecurity, and conflict. Thus, no basic changes can be effectively summarized in the area of "professional handling of the parent."

Habilitation

The techniques of habilitation of deaf youngsters have multiplied, become sophisticated and require ever more training for optimal implementation. Hearing aids and their early usage have become technically refined and new improvements continue in their performance because of advanced knowledge of psychoacoustics. Lipreading and speech therapy are undergoing innovative changes. Education for deaf youngsters has shown tremendous strides. Nevertheless, yearly publications deplore the overall achievement of a large percentage of deaf children grown up, despite clearly normal potential (Schlesinger and Meadow, 1971).

However, the area of habilitation is even more beset with intangible feelings whose behavioral concomitants clearly interfere with the optimal dissemination of information and utilization of facts for the deaf child.

In the Early Years: Parents clearly yearn for the eventual "total normalcy" of their hearing impaired child, a normalcy which is to include normal speech and pefect lipreading facility, despite innumerable stories and research of the relative infrequency of such results. The professionals, either by ignorance, but much more frequently by the above-mentioned reluctance to give out "bad news," encourage the unrealistic and rosy picture. Parents also frequently relate that they have encounters with experts who vigorously espouse conflicting advice about myriad matters: which type of hearing aid? one or two? or none? at what age?; lipreading only, auditory training only, a combination of both with speech training, or the addition of sign language and fingerspelling to speech. Each advice is either given or perceived by the parents in Old Testament admonitions:

> "Thou shalt always talk to the deaf child, thou shalt never gesture to the deaf child, thou shalt always praise him and never be angry with him, thou shalt treat him as if he were not deaf, thy lips shall be at eye level for the child, thou and only thou are

responsible for his ability to wear his hearing aid and if thou does not follow our heed he will be lost to thee." (Schlesinger, 1968)

We have encountered a number of patients who were either relatively successful or unsuccessful with any or all of the above techniques. What appears to be of increasing importance is the quality of the advice rather than its pure content. This quality has been further subdivided by Ross (1964) into human and professional qualities. The human qualities frequently exist in individuals otherwise untrained in the behavioral sciences such as interest, acceptance, flexibility and sensitivity to both the parent's and the child's needs. These human qualities can also be developed throughout the course of careful training in therapy or counseling and must be further supplemented by

> "the professional attributes of objectivity, confidence, and knowledge, as well as the technical skill of interviewing. The professional helper must be familiar with available community resources, he must know when and how to obtain consultation, and he must have a realistic appraisal of the limits of his competence." (Ross, p. 75)

It is indeed difficult, but may be imperative, to find a professional with adequate knowledge of the specific audiological, speech and hearing aid requirements for the deaf child, who also combines the above qualities, to impart his knowledge with skill and support to the parents.

The Later Years: Habilitation in the later childhood years is primarily an educational function. The physician further recedes in importance, although he continues to fulfill two important roles. One, conductive deafness is frequently intermittently superimposed on the sensorineural deafness, and conductive deafness is amenable to medical cure. Furthermore, the physician is frequently called upon to act as support to parents who remain perplexed about the progress of their deaf child.

Let me terminate with a short summary of some of our own work which I think poignantly traces the intricate network of research findings, clinical findings, and the subsequent sharing of impressions with parents in an attempt to thank them for their patient collaboration with our work, as well as to provide requested alleviation of their anxieties. Our clinical findings increasingly tended to point to the fact that the ubiquitous controversy between "oralism" and "manualism" was in itself detrimental both to child and parent and when alleviated permitted spurts of growth, no matter what "methodology" remained the preferred one. On the other hand, our research continued to gather evidence indicating that youngsters whose parents used speech and sign language at an early age (8-21 months) and did so joyfully and without the ubiquitous conflict had a distinct linguistic, speech, and lipreading advantage. How to combine these two findings which are complementary and yet not identical in a helpful way to parents caused (and still causes) a unique therapeutic experience. Some parents are so conflicted in the either/or controversy that a relaxation of the conflict

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was of utmost importance, and yet feelings engendered by the conflict were so intense that they precluded receptivity to our findings, which in turn might have reduced the conflict without changing the content of the opinion.

Some parents remain unaware how pervasive the effects of their conflict is on the child's growth, whereas others came with no conflict but with insufficient knowledge about any aspects of language acquisition of their children, be it oral, manual, or a combination.

All of these parents have greatly contributed to our increasing knowledge about the prevention, diagnosis, and habilitation of deafness. We thank them and hope that this publication will help others develop an interest in this rare but important childhood disorder.

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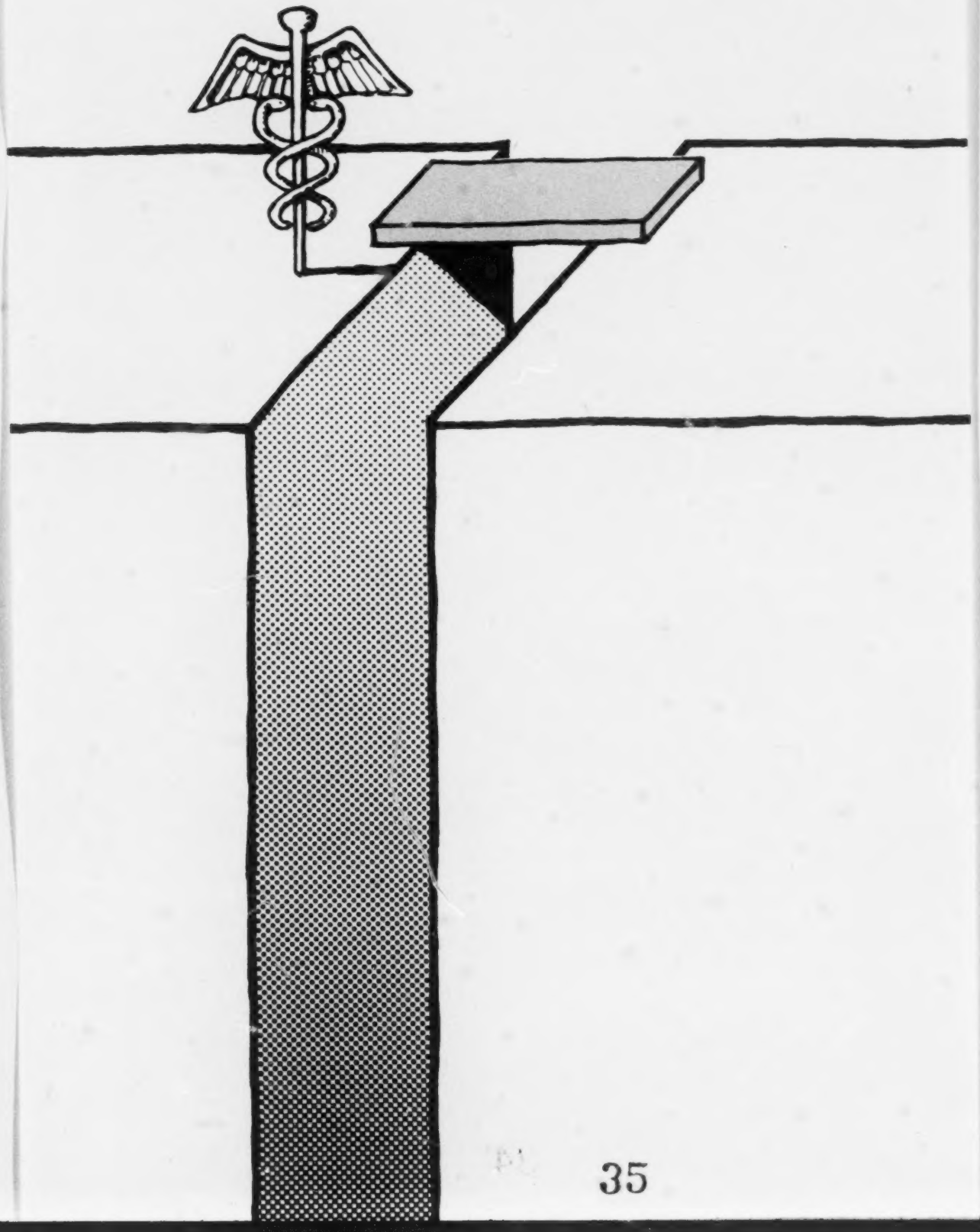
Prevention



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GENETIC AND ENVIRONMENTAL FACTORS IN DEAFNESS

Early total deafness occurs in the population at the rate of about one per thousand. We showed that about one-half of this deafness is hereditary, with the other one-half caused by early environmental traumata. We found that deaf persons are marrying more frequently than they did in earlier decades, although their marriage rate is still not up to that of the general population. More than 90 percent of all deaf persons are the offspring of two hearing parents, but whether they themselves had deaf parents or hearing parents, deaf persons seemed to marry at about the same rate. In the case of deaf individuals who were born deaf or who have deaf relatives, the percentage of deaf brothers and sisters is about 24 percent. The fertility rate of deaf mothers is somewhat below that of the general population, but again much higher than it was relative to the general population in earlier decades.

Statistically, only five to nine percent of deaf women are married to men with normal hearing. The remainder have husbands who have been deaf since birth or early childhood. In marriages where both partners were born deaf, about 30 percent result in deaf children. In those where only one partner was born deaf, from 14 to 41 percent result in deaf children. Of all the children born to all the deaf subjects in New York, about 10 percent are themselves deaf. This is very similar to the figure pointed out more than 60 years ago in the well known study by Fay. Finally, it is important to point out that, of the parents of deaf persons in New York, the marriage rate between cousins was almost nine percent—much higher than the one-half to one percent figure for cousin marriages in the general population.

Genetics of Deafness

Regarding the genetics of deafness itself, further details can be found in a number of places, particularly in an article by G.R. Fraser or in the chapter by Diane Sank in our own study. I would like to summarize the facts here: According to Fraser, 46 percent of early total deafness is acquired, the remainder is genetic. The acquired cases include six percent prenatally (including the rubella cases), 10 percent perinatally (including prematurity, Rh factor, etc.), and 30 percent postnatally (including, particularly, meningitis).

The genetic cases include almost 40 percent because of recessive inheritance, about 12 percent owing to dominant inheritance, and about one and one-half percent sex-linked. The recessive cases include about one-fifth with retinitis pigmentosa, the condition in which the deafness is followed by progressive loss of vision (particularly peripheral vision), a small number with goiter, a small number with a rare syndrome including abnormal electrocardiogram and cardiac problems, and all of the other recessive cases, by far most of them being caused by one of perhaps 30 or 40 different recessive genes. This is important because an individual may carry a recessive gene for deafness and marry someone who carries a different recessive gene for deafness and their children will all be hearing. It is only if they both carry the same recessive gene that the children will be deaf and, since there seem to be 30 or 40 genes, this is a rather rare occurrence. Just to mention the dominant cases, about one-fifth of them are associated with pigmentary anomalies. Chief among these is the Waardenburg syndrome, in which deafness is accompanied by a white streak in the hair, the eyes are set somewhat wide apart and are often of different colors. You will remember, of course, that in a dominant condition, if one parent has the condition, a child has a 50 percent chance of having it, whereas in a recessive condition it is necessary for both parents to be carriers of the condition, although neither may have the condition themselves.

Figures Corroborated

Now, the figures obtained in our own New York State study by Sank corroborate very well those figures collected by Fraser. Again, about one-half of all deafness seems to be sporadic, the other one-half includes about 4/5 recessive and 1/5 dominant. A further finding of Sank, which came out of some of the twin studies she did while working in our group, was that often two individuals in the same family might have different degrees of deafness. There might be a partial hearing loss in one and deafness in the other, even though because of the same genetic factors. And, finally, it may very well be—from some of the results obtained—that certain individuals are more susceptible from a genetic point of view to becoming deaf as a result, let us say, of meningitis than of others. In other words, there is always an interaction between genetic and environmental factors, and in any case it is only a question of which exerts the greater influence.

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PREVENTION OF DEAFNESS

The topic of prevention of deafness can be approached in several ways. I have chosen to look at the types of deafness classified as to their cause. We will then examine what can be done to prevent deafness in each causal group. Traditionally, we can state that three main causal groups of deafness exist. The first is that of acquired deafness which consists of infection, prematurity, sound trauma, ototoxicity, etc. The second is that of genetic deafness which can be either dominantly transmitted or recessively transmitted. The third category is that of unknown or idiopathic deafness which is seen all too frequently.

The percentages of different types of deafness which have been found in our clinic population of about 360 patients show that approximately 21 percent are genetic, about 39 percent are acquired, and the remaining 40 percent are unknown.

Much is already known about what can be done for the eradication of almost all types of acquired deafness. The most spectacular and far-reaching advance in recent years has been the development of the rubella vaccine, which is effective in most cases. However, there are some instances in which the vaccine has been reported as functioning improperly. It has been the practice in some institutions, especially in Scandinavia, that the expectant mother has a blood titer drawn at the beginning of her pregnancy and again at the end of the third month of pregnancy. If the rubella titer has become elevated during that time, she is advised of the possibility that she may have had a sub-clinical infection of rubella. A decision can be made at that time as to whether or not the pregnancy should be continued.

Another area in which a tremendous advance has been made in the prevention of acquired deafness is that of infectious disease. Today it is only the rare case that is seen with deafness secondary to bilateral labyrinthitis, that is to say, infection of the inner ear. However, a large number of children and adults still develop hearing loss, in the moderate to severe category, secondary to middle ear infection. It is strongly felt that much of this disease can be prevented. One group of the population which is especially prone to middle ear infections are those who have cleft palates. Some studies have indicated that in a group of cleft palates which receive no otological care, as many as 25 percent will have serious tympanomastoiditis with the resultant moderate to severe hearing

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loss. Many pediatric otologists today have done small procedures; that is, the placing of polyethylene tubes, which have aborted most of the serious sequelae of tympanomastoiditis in this group of patients.

Improper Facilities

We also know our large urban slum areas contain innumerable children with bilateral serous or mucoid otitis media. This disease results in only a moderate hearing loss but it has grave sequelae later, in the ability of the child to use language and in the development of his entire verbal intelligence. Technology is now available to care for most disease in these children. However, our society today does not allow us to establish—especially in the ghetto areas of our large cities—the proper diagnostic screening facilities. Then, too, if the children could be screened, medical facilities are not available (at least not in my city) to take care of what we know we would find. Although this is not a problem of profound deafness, it has a very real place in our total consideration of the prevention of hearing handicaps. I am talking now about the group of hearing handicapped children with delayed, retarded language and verbal ability who live in culturally and nutritionally deprived ghetto areas. More research work is not necessary in this area; merely apply what we have and we could most likely improve the verbal and language I.Q. of a significantly large segment of our ghetto population.

Prematurity is a more complex problem of preventive medicine. There is little understanding as to the exact mechanics of communicative disorders arising from prematurity. Some premature infants are noted to have peripheral disease in the cochlea or the VIIIth nerve, whereas others may have central nervous system disease in terms of auditory agnosia. Some of the children will have combinations of both. The prevention of hearing loss in prematurity has to do with the prevention of prematurity in general.

A factor known to have some importance is that of ototoxic medication. The use of such medication in premature infants should be closely regulated so that the infant does not have a toxic dose. Unfortunately, ototoxic antibiotics are still widely used throughout the medical profession. Approximately 10 percent of our adult patients with acquired deafness have become totally deaf due to the use of ototoxic antibiotics. In most cases, much of this deafness could be prevented. The prevention could be accomplished either by the use of another antibiotic, by monitoring the blood levels, by using the proper levels of antibiotics and/or by using artificial kidneys and other dialyzing techniques so that the blood concentration of the ototoxic antibiotic does not develop. Prevention here is really the education of physicians using antibiotics as to the catastrophic sequelae of sudden acquired deafness secondary to ototoxic drugs.

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Preventing Genetic Deafness

The prevention of genetic deafness is at the present time a very passive type of problem. There are two main types of genetic transmission of deafness. The most commonly noted but probably not the most common in existence is the so-called dominant transmission. Only one parent has to be affected with one gene. Half of the children from this parent will be affected, more or less, with deafness. The parent can be advised that he or she is carrying the gene and that probably the children will have the disease. Recessive transmission is somewhat more complicated. Each parent must have one gene and the child, in order to be deaf, must have a gene from each parent. The probability of each birth being so affected is one in four. Again, our best weapon in preventing deafness here is advising the parents that they are carrying the gene and that there is a very real probability that each child has a one out of four chance of being affected with the deafness gene.

The unknown category, which in our series accounts for some 40 percent of the patients, is by and large the most interesting. When geneticists look at the distribution of etiological factors of deafness, they break it down somewhat differently than we, as clinicians, do. They have no unknowns and put their unknowns in the recessive category. I suspect that the truth lies somewhere between the clinical data and the genetic data. However, the point to be made here is that many of our unknowns are patients who are carrying deafness genes. The most conservative estimate as to the number of recessive deafness genes in the population is that one out of every four persons has a gene for deafness. This accounts for much of the spontaneous hearing loss which is seen. It has been our practice to advise a parent who has one deaf child for which there is no known cause that a possibility exists the child has a genetic hearing loss. We cannot rule this out. In many instances where the first child was born deaf, the parents were not advised of the possibility that they could have another deaf child, hence, they have produced a second and sometimes a third and fourth deaf child.

Controlling Acquired Deafness

The picture which I have presented so far is not too unpleasant. However, in many ways I feel ashamed to be an otolaryngologist in this day and age when it comes to the problem of inner ear deafness. All of the things I have talked about have been relatively passive ways of prevention with little understanding of the disease processes. There has either been a vaccine, an abortion, or prevention of a birth. I feel confident that soon we will be able to control most deafness which is acquired and that in the group of unknowns we will be able to identify many more acquired types of deafness than we can now. These types may be caused by other RNA viruses, such as rubella, which heretofore have not been directly associated with deafness.

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However, there will always be a large number of persons who are carrying genes for deafness. These genes are thoroughly mixed in the population, and people also have the ability to mutate a gene for deafness. The control of deafness due to genetic factors depends on a biochemical understanding of the enzyme defect which is associated with specific genetic abnormalities. With the exception of Refsum's disease, a very rare and uncommon disease, we do not have the enzymatic biochemistry of any genetic disease of deafness worked out. All other major diseases have this type of information. We can now in many cases rationally treat, or perhaps even reverse, disease processes. For deafness we can do none of this.

There is a great deficit of knowledge, at the cellular level, of the enzymes and their interaction with abnormal genes which cause deafness. I close this brief discussion with a plea that resources be used for understanding the problems of the ear at the cellular level. I feel that much excellent work has been done on the physiology, anatomy, and ultrastructure of the inner ear and the middle ear. The time is now quite ripe for a concerted attack on the cellular mechanics of the ear, so that we can begin to understand the cellular processes involved in the death of the hair cells, and perhaps find ways of reversing this and also of stopping the onslaught of genetic disease within the ear.

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HEREDITARY DEAFNESS: AN APPROACH TO DIAGNOSIS, PREVENTION, AND TREATMENT

The laws of genetics in human disease are becoming increasingly relevant as our understanding of the basic biochemical mechanism of inheritance increases by leaps and bounds and as the possibility for prevention through genetic counseling and prenatal diagnosis enables us to contend more definitively with the possibility of elimination of some genetic diseases in the population. Fortunately, these two approaches often are supported by therapy for individuals already affected, where prevention obviously is not possible.

Genetics is particularly relevant to the problem of deafness in regards to all three aspects–diagnosis, prevention and treatment. There is no need to emphasize the significance of hearing loss as a health problem for this particular audience. It should be noted, however, that genetic causes probably represent the explanation for approximately 50 percent of individuals with congenital deafness.[1]

For the past five years we have been engaged in a study supported by the National Institutes of Health, in which we have approached the problem of hereditary deafness from a number of points of view. Our study involves the cooperation of specialists in medicine, otolaryngology, audiology, radiology, social service, and psychology. Today we should like to describe a typical case history as it evolves from its initial presentation to our study group through its ramifications for prevention through genetic counseling, and possible treatment through surgical manipulation.

Initially, however, we should like to outline the types of hereditary deafness which have been described, and explain the major modes of inheritance which are recognized.

Hereditary deafness has been conveniently classified into two major categories:[2]

(1) Hearing loss associated with other abnormalities

(2) Hearing loss unassociated with other abnormalities

Examples of such syndromic diseases are Usher's syndrome, where hereditary deafness is associated with blindness, and Alport's syndrome, where hereditary deafness is associated with nephritis. (Fig. 1) The convenience of this syndromic approach is obvious. If one can definitely associate deafness with a non-hearing abnormality, diagnosis of a genetic etiology may be possible even in the absence of a positive family history of deafness.

The second group (Fig. 2) is less well-defined. In this group, however, an hereditary etiology can still be suspected if there are multiple affected relatives in the family, if the audiogram shows a diagnostic pattern, if parental consanguinity is present, if minor audiologic abnormalities can be detected in close relatives, or if it is possible to link the pedigree to a roster of familiies known to have hereditary deafness.

The major modes of inheritance are dominant, recessive, and X-linked. A pedigree demonstrating dominantly inherited low-frequency hearing loss is shown in Figure 3.[3] The black squares denote affected males and the black circles affected females. As one can see, the mode of inheritance passes directly from one generation to the next, without regard to sex and without the omission of any generation. In other words, for every affected individual there is an affected parent. In this particular kindred, we were able to further define the hereditary deafness by the typical low-frequency hearing loss pattern. (Fig. 4)

Recessive Loss

A typical pedigree of recessive hearing loss is shown in Figure 5.[4] While consanguinity is not invariably found with rare recessive traits, it can often be the only clue that one is, in fact, dealing with a recessive trait. The double lines represent consanguinous marriages and, as is readily seen, the affected individuals, again shown in black, do not appear in repetitive pattern from generation to generation but, rather, skip about the pedigree, although again there is no relationship to the sex of the individual. In this type, an affected individual need not have an affected parent. The pedigree of another family with recessively inherited hearing loss showed similar characteristics (Fig. 6). With these two families we have been able to demonstrate a different composite audiogram, suggesting that, while both families have recessively inherited hearing loss, the type of hearing loss is different between the two. (Fig. 7)

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The next illustration (Fig. 8) shows a pedigree in which deafness is transmitted as an X-linked trait.[5] Hemophilia, muscular dystrophy, and color blindness are familiar examples of other genetic traits that show a similar pattern of inheritance. The abnormal gene is located on the X chromosome, and is fully expressed in affected males, because males carry only one X chromosome along with a very much smaller Y chromosome. Females, on the other hand, have two X chromosomes and ordinarily would not be expected to manifest an X-linked trait unless they carried two copies of the abnormal gene, one on each chromosome. Finally, males inherit their Y chromosome from their father; otherwise they would not be males. Consequently, the single X chromosome that males carry invariably comes from the mother. In the present pedigree we see that all of the affected individuals are males (squares), and that they are all linked to each other through female relatives. This is the pedigree pattern one would expect with an X-linked trait. On the average, we would expect one-half of the sons of a carrier mother to be affected and one-half of her daughters to be carriers. An affected male would have normal sons, but all of his daughters would be carriers. Finally, normal males in the family do not carry the gene and, therefore, would have no risk of transmitting the trait to their children.

Turning now to our typical case history and utilizing the family shown in Figure 8, we will demonstrate how our study functions. The procedure in regard to initial contact with the family, pedigree research, clinic evaluation, and hospitalization will be described.

Every patient seeking hearing evaluation at the Bill Wilkerson Hearing and Speech Center is requested to complete the Family Questionnaire before his audiologic appointment. (Figs. 9 and 10) More than 1,200 questionnaires, containing at least 30 names per questionnaire, have been collected during the past five years.

A glance at the questionnaire will indicate its usefulness in choosing patients for intensive pedigree study. Page one of the questionnaire gives a brief medical history with clues to indicate whether deafness of the individual may have been caused by accident, illness, or drugs. This page also indicates whether there are signs or symptoms of a known syndrome. The other pages of the questionnaire contain the family tree with a history of deafness for ancestors and descendants. We look at these pages to choose families with a strong history of hereditary hearing loss and to learn whether the same surnames occur in both the paternal and maternal sides of the family, indicating a possible common ancestor.

The family in this example became known to us when a three-year-old boy was admitted to the Bill Wilkerson Center for hearing evaluation. (Fig. 11) The examining audiologist was alert to the history of deafness in other generations of this family and referred him to the Hereditary Deafness Clinic. Because she was aware of the three-generation history of deafness in her family, the child's mother was conscious of the advisability of doing everything possible to help with a

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study such as ours. She interested other members of the family, who lived at a considerable distance, and they, too, visited our clinic.

Obtaining Data

I should like to comment about some of the dynamics of obtaining pedigree data. In practically every family studied, it has been apparent that we need to enlist the cooperation of one or two actively interested family members. They are the ones who will gain the confidence of their relatives and make the effort to seek out pedigree data. This may mean finding the missing family Bible or going to the cemetery to locate dates and places of birth. Also, research in hereditary deafness can imply triple threats to some families. Research, to many persons, suggests experimentation. Deafness of children, for complex reasons, is an emotionally loaded subject for the parents and, when coupled with the word "hereditary," is often unacceptable. Thorough study of kindreds with familial deafness, therefore, requires a sensitivity in approach, the ability to present facts without causing needless pain, and, most important, a genuine acceptance of these families that geneticists have estimated that every person carries from three to four abnormal genes, which, if present in double dose, would cause a serious abnormality such as deafness, blindness, or mental retardation. Knowing what one of these genes is can be an advantage for an individual or a family.

Most persons require much support in the process of revealing familial data which, however innocuous it may sound to the interviewer, may have stigmatizing implications for the family. This was pointed out to us clearly at the beginning of our study when certain members of a family reacted with hostility to our interviewing their mother who had always gone to great trouble to conceal the white streak in her hair. This almost unnoticeable sign, coupled with slightly different colored eyes, was indicative to us that she had some of the signs of Waardenburg's syndrome although she had escaped the deafness. One other point of importance, of which we have become aware, is the fact that persons, having produced a deaf child, must find some explanation acceptable to themselves that leads eventually to a state resembling emotional equanimity. Close inquiry into this situation can arouse hidden anxieties. One of the requirements, then, would be to help such persons regain their emotional equilibrium. Hopefully, they may obtain a healthier and sounder perspective, through the experience of having faced their inherited traits with persons who are uncritical and with whom a therapeutic relationship can develop. When accompanied by an insight into the frequency of abnormal genes in the general population, a genetic diagnosis can relieve feelings of guilt, shame, and uncertainty and allow the parents to plan for the future in a rational manner.

The child in this example, selected for evaluation on the basis of pedigree information obtained through the Family Questionnaire, was evaluated by our specialists. He received a complete medical history and

physical examination, otolaryngologic examination, and intensive audiometric testing. The examination of individuals, whose age and hearing level permit, includes pure tone air and bone thresholds, speech reception thresholds, discrimination scores, SISI tests, Bekesy tracings, and tympanometry. Some of our patients may receive special radiologic evaluations and other out-patient consultations as needed. In certain families, affected individuals are hospitalized in the Vanderbilt Hospital Clinical Research Center for more intensive study. Other family members receive detailed evaluations, also.

In the particular family under consideration, studies were accomplished on seemingly non-affected individuals. Further studies in this family revealed that the carrier state may be recognized by its audiograms (Fig. 12). These females have some hearing loss, demonstrable by testing, although superficially they appear to function normally. Through detailed audiologic testing, we may be able to recognize those females who can transmit the defect, providing us with greater potential for accurate genetic counseling in a given family.

Our three-year-old boy in this family was hospitalized, but no significant findings were noted, other than hearing loss. Our quesionnaire had revealed that the family contained 10 deaf males in three generations in a pattern consistent with X-linked inheritance. This deafness, as was previously mentioned, may be transmitted by the mother to male offspring. Daughters of affected or carrier individuals may also be carriers and transmit the disorder to their sons.

Because of the likelihood of his having a similar defect producing deafness, a 51-year-old great uncle of this child was also hospitalized. Exploratory surgery was performed because the audiogram showed a conductive component to the hearing loss. At surgery, it was observed that there was fixation of the stapes footplate. Mobilization of the stapes was attempted at this time, but a profuse flow of clear fluid, assumed to be perilymph, ensued, filling the middle ear. This was the result of a patent aqueduct. The surgical procedure was terminated at that point and, since mobilization of the stapes was not feasible, no improvement in hearing was accomplished.

Subsequent Developments

Having described the diagnostic approach to a family with hereditary deafness, I should now like to discuss subsequent developments resulting from our study of this family, genetic counseling aspects relative to prevention, and therapeutic possibilities. In this particular family, the finding at surgery in one individual led to the recognition that similar gushers, previously described elsewhere, no doubt represented the same genetic entity. Two similar patients elsewhere have had narrowing of the patent aqueduct accomplished surgically, followed by mobilization of the stapes.[6] In addition, we have subsequently found in our clinic two more families with X-linked inheritance patterns and similar anatomical

abnormalities. It would now appear that we have a specific genetic entity, recognizable by its inheritance pattern and anatomical abnormalities, which produces deafness.

The first question asked by such a family would be, "Why is my child deaf?" In a family where there is an affected male child and where the pedigree has revealed multiple instances of affected males, in the same and/or other generations of the family, without affected females, one can be certain that one is dealing with X-linked inheritance, and can explain this to the family.

The next question such a family would ask would be, "Should we have further children?" The chances of having another affected male child would be 50 percent. In addition, the chances of having a female child who could pass on to 50 percent of her male children the same defect would be 50 percent. With this knowledge, the parents can make a rational decision about whether to have more children or not. While prenatal diagnosis is not yet feasible, in this particular disease prenatal sex determination could permit the identification of those children (males), who are at risk of being deaf.

The next question would be, "Can anything be done for our affected male child?" Unfortunately, our knowledge in many instances is not far enough advanced to allow us to give a positive answer. However, in the family shown here, we do believe that the defect may be amenable to surgical repair. As mentioned, it has been found at exploratory surgery that there is persistence of the cochlear aqueduct associated with fixation of the stapes and that by appropriate narrowing of this aqueduct, with mobilization of the stapes, possible improvement in hearing may result. We are planning to attempt several such trial surgical procedures on affected members of this kindred in the near future.

Thus, our three-year-old child has led us through an expanding recognition towards a new genetic entity, the inheritance of which is understood and for which a surgical procedure may be developed to help affected individuals.

This brief sketch of a typical case history demonstrates the feasibility of the genetic approach to deafness in terms of diagnosis, prevention, and treatment. The understanding of genetic modes of inheritance and the recognition of syndromic genetic deafness does not necessarily require extensive medical education. We hope to institute a program of training for audiologists to include a basic understanding, such as outlined here, in order that they who often form the initial contact with individuals with hereditary deafness may have a greater ability for the recognition of such difficulty and ultimately participate in the genetic counseling of affected kindreds.

In summary, then, the recognition of hereditary deafness involves an understanding as to the incidence of hereditary deafness, the types of hereditary deafness which one may see, and the genetic modes of inheritance. At the present time, prevention involves primarily genetic

counseling, although, as our knowledge expands in other areas, other forms of prevention may well become feasible. Finally, it is through the expansion of our understanding of hereditary deafness that therapeutic possibilities, such as the example given above, will hopefully ensue.

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Fig. 1

PARTIAL LISTING OF SYNDROMES WITH HEARING LOSS

Hereditary Deafness with External-Ear Malformations:
Dominant Preauricular Pits and Neural Hearing Loss
Recessive Malformed Low-Set Ears and Conductive Hearing Loss

Hereditary Deafness with Integumentary Disease:
*Waardenburg's Disease
*Hereditary Piebaldness and Congenital Deafness
Sex-Linked Pigmentary Abnormalities and Congenital Deafness

Hereditary Deafness Associated with Eye Disease:
Dominant Myopia, Hearing Loss, Peripheral Neuropathy and Skeletal Abnormalities
*Recessive Retinitis Pigmentosa with Congenital Deafness (Usher's Disease)

Hereditary Deafness Associated with Nervous-System Disease:
Dominant Sensory Radicular Neuropathy
Recessive Deafness, Mental Deficiency, Ataxia and Hypogonadism (Richards-Rundel Disease)

Hereditary Deafness Associated with Skeletal Disease:
Craniofacial Dysostosis (Crouzon's Disease)
*Mandibulofacial Dysostosis (Treacher-Collins Disease)

Hereditary Deafness Associated with Renal Disease:
*Dominant Nephritis with Hearing Loss (Alport's Disease)
Recessive Renal, Genital and Middle-Ear Anomalies

Hereditary Deafness Associated with Other Abnormalities:
Recessive Goiter and Deafness (Pendred's Disease)
*Recessive Heart Disease and Deafness (Jervell and Lange-Nielsen Disease)

* Seen in Hereditary Deafness Clinic of "Experimental Studies in Hereditary Deafness"

From: Konigsmark, B. W., M.D., "Hereditary Deafness in Man", New England Journal of Medicine, 281: 713-720, 774-778, 827-832 (Sept. 25), (Oct. 2), (Oct. 9), 1969.

Hereditary Deafness with No Associated Abnormalities

*Dominant Congenital Severe Deafness
Dominant Progressive Nerve Deafness
Dominant Unilateral Deafness
*Dominant Low-Frequency Hearing Loss
Dominant Mid-Frequency Hearing Loss
*Otosclerosis
*Recessive Congenital Severe Deafness
Recessive Early-Onset Neural Deafness
*Recessive Congenital Moderate Hearing Loss
Sex-Linked Congenital Neural Deafness
Sex-Linked Early-Onset Neural Deafness
Sex-Linked Moderate Hearing Loss

* Seen in Hereditary Deafness Clinic of "Experimental Studies in Hereditary Deafness"

From: Konigsmark, B. W., M.D., New England Journal of Medicine, 281: 713-720, 774-778, 827-832, (Sept. 25), (Oct. 2), (Oct. 9), 1969.

Fig. 2

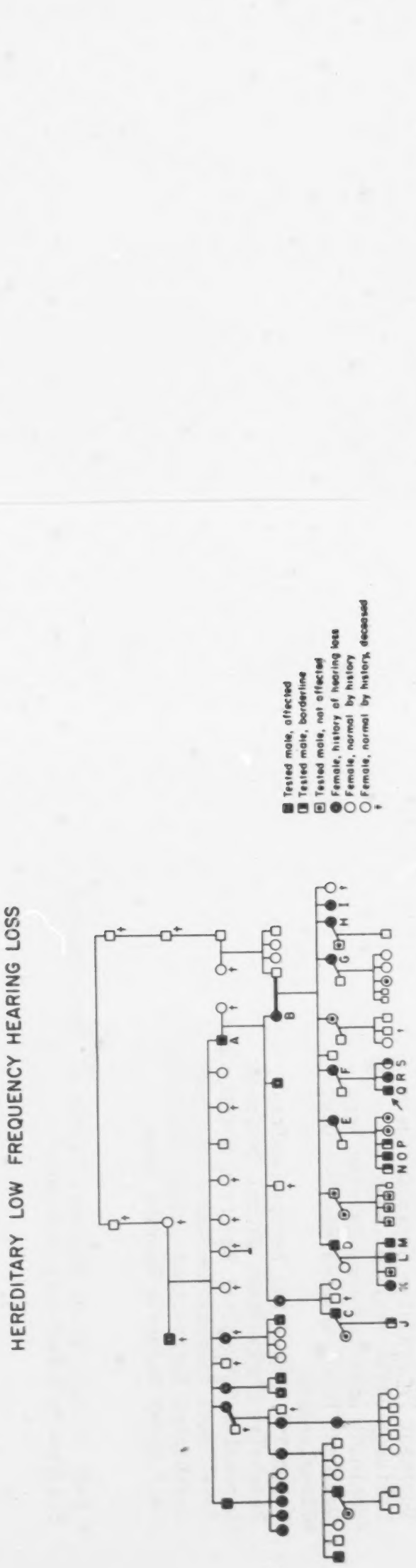

Fig. 3 Pedigree of kindred with autosomal, dominantly inherited hearing loss showing 21 individuals examined and found to be affected; four individuals examined and found to have borderline low-frequency hearing loss; and 13 individuals, not examined, with history of hearing loss.

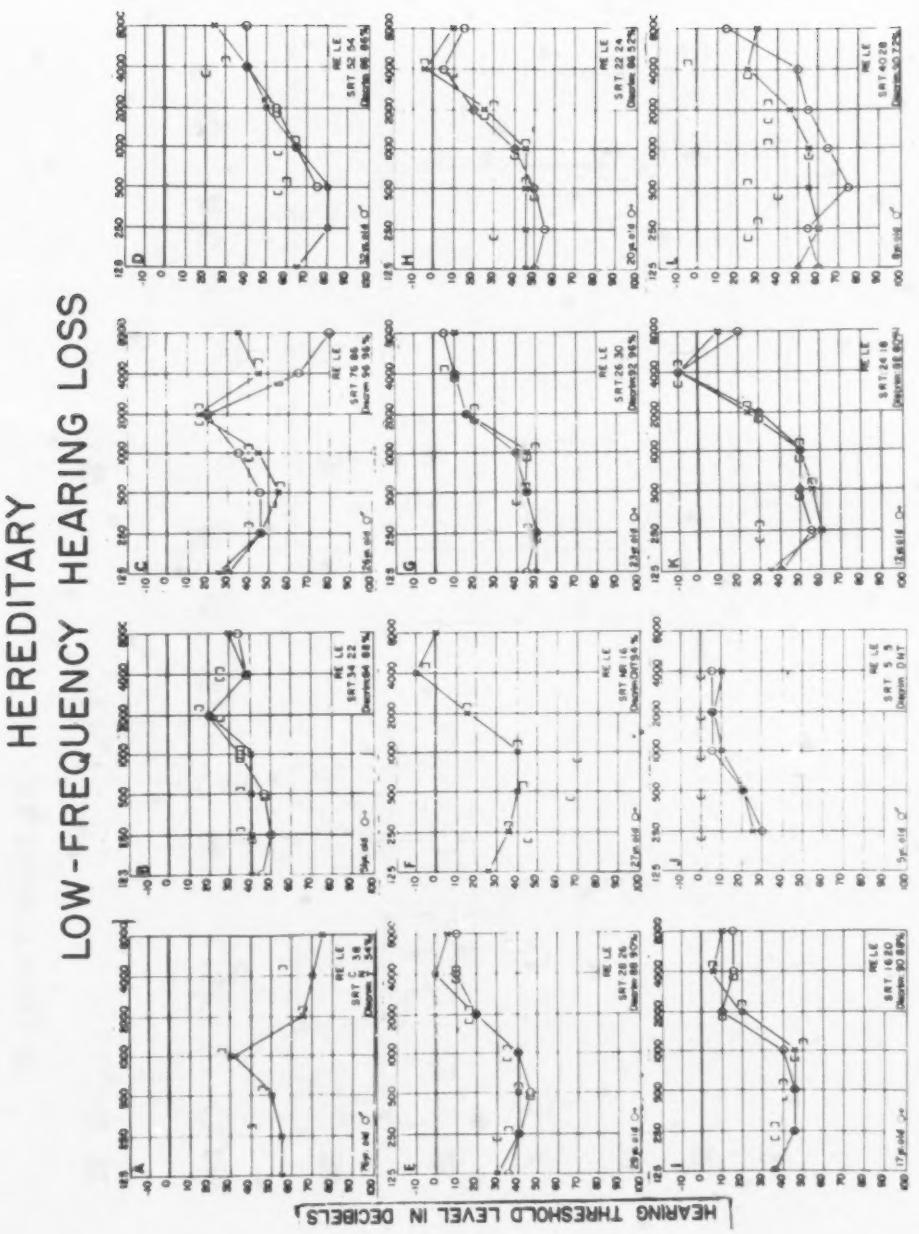

Fig. 4 Audiograms of 12 closely related individuals, ranging in age from five to 76 years, affected with dominantly inherited low-frequency hearing loss.

52[illegible]

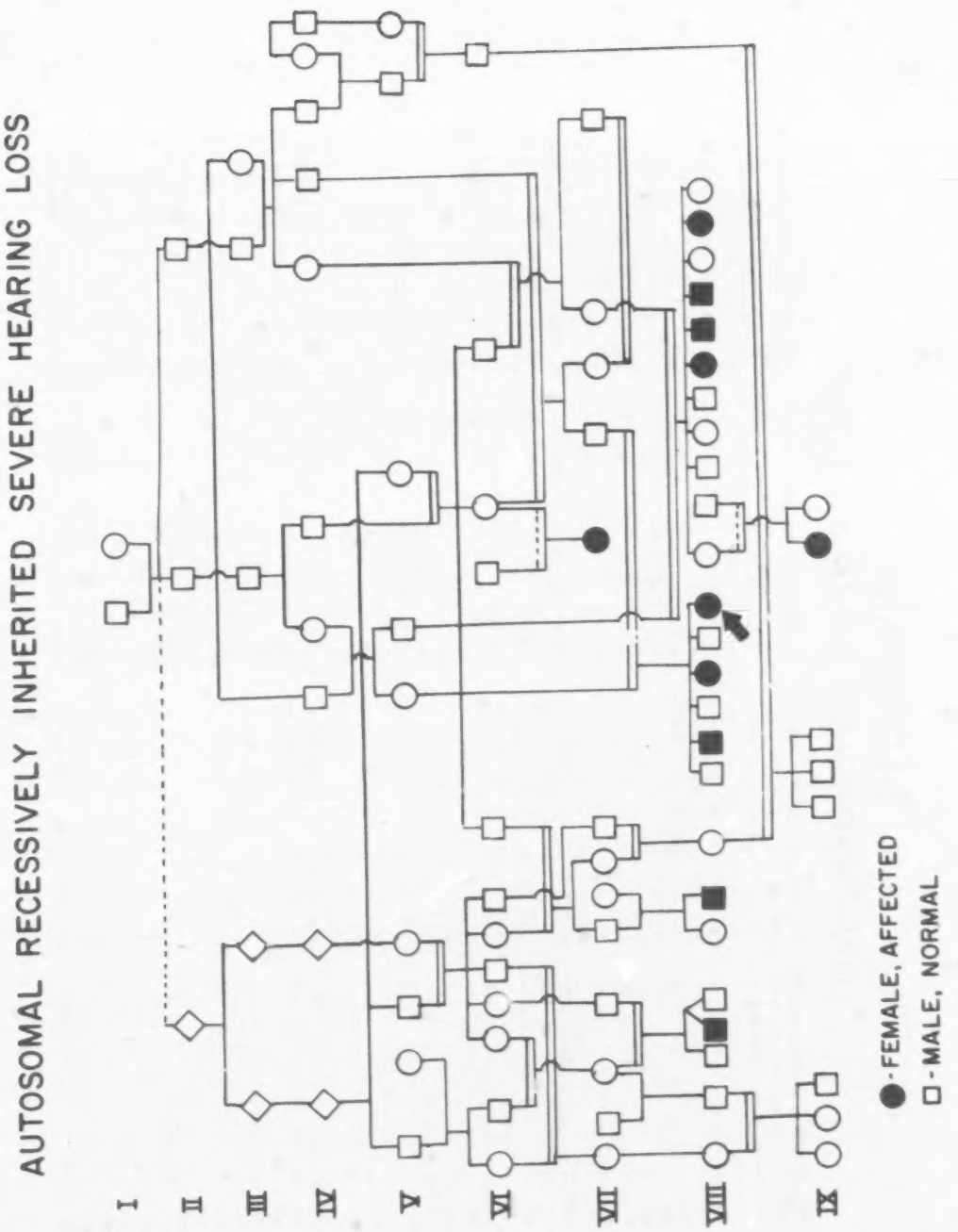

Fig. 5 Pedigree of family with recessively inherited deafness showing 11 affected offspring of consanguinous matings.

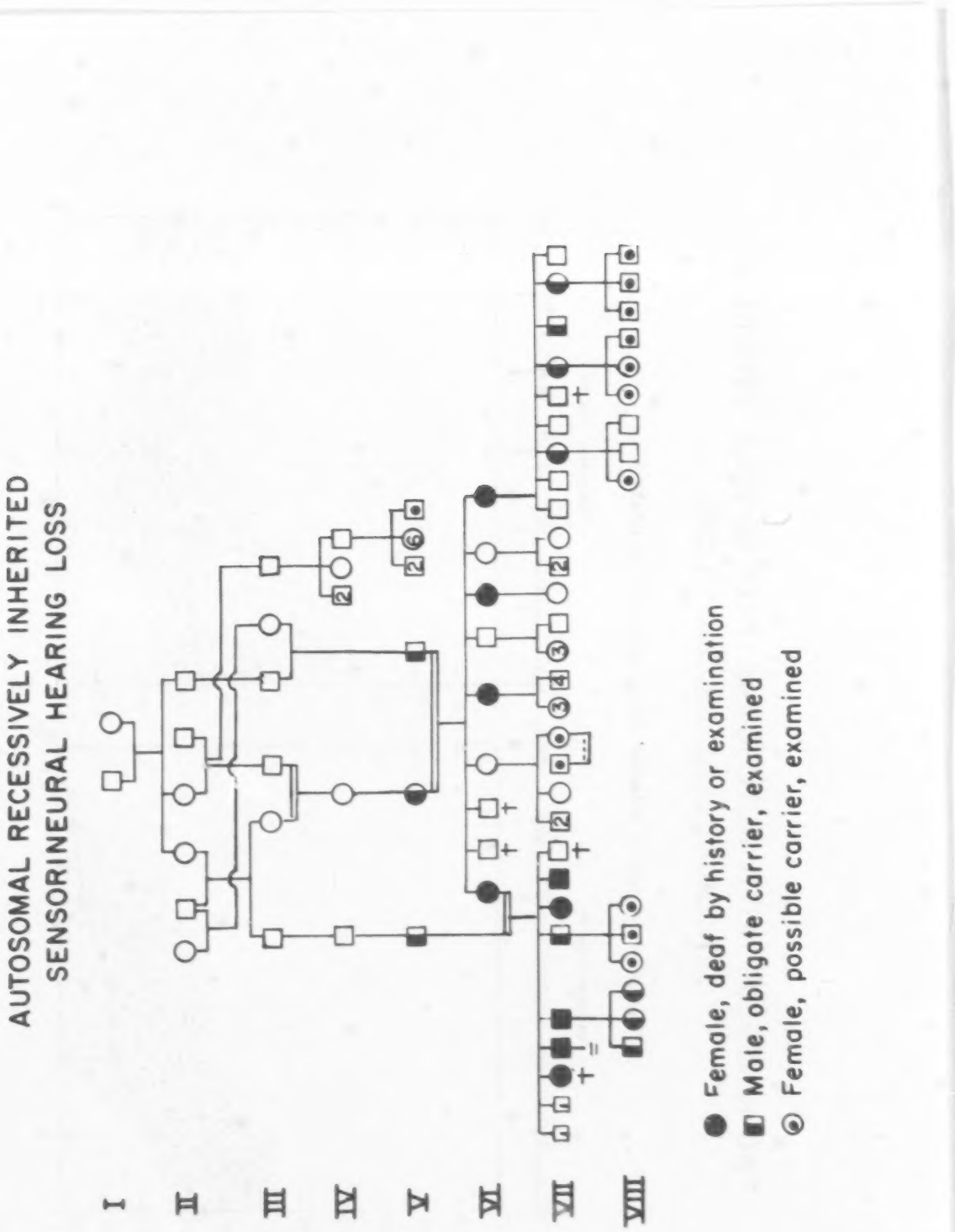

Fig. 6 Pedigree of family with recessively inherited hearing loss showing nine affected individuals occurring in inbred sibships.

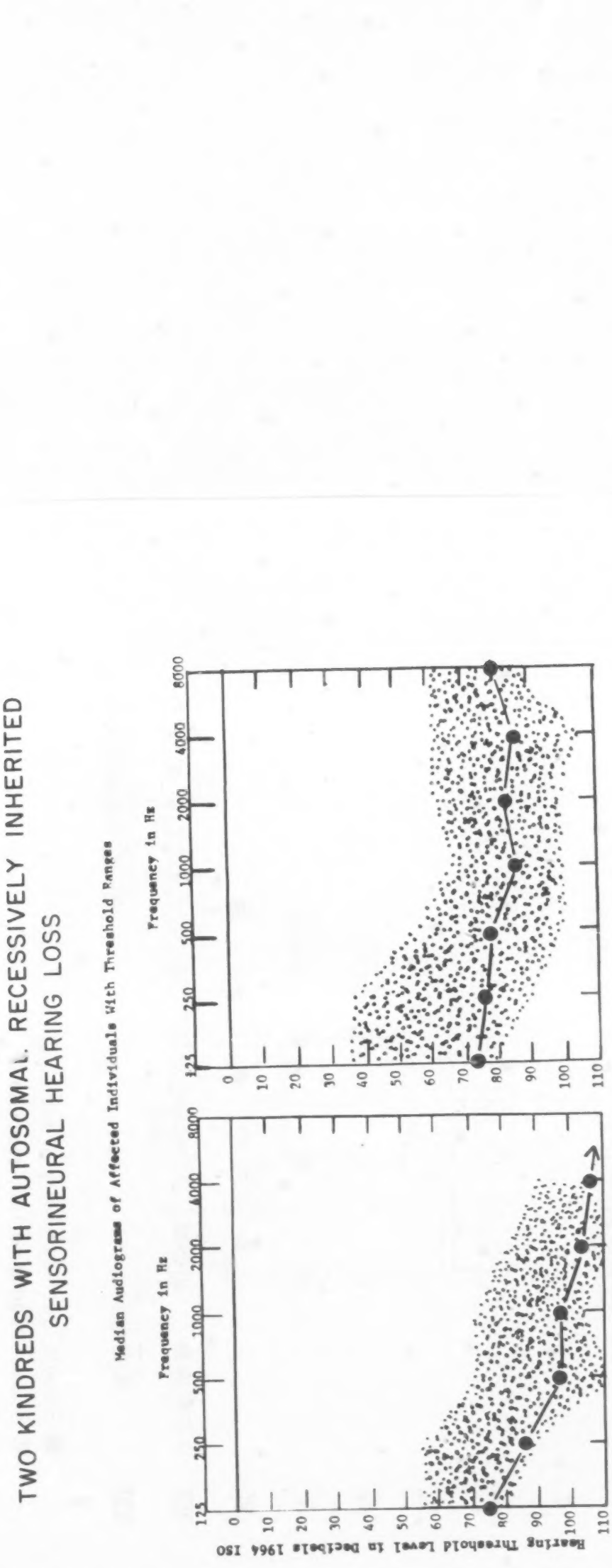

Fig. 7 Composite audiograms of individuals affected with recessively inherited hearing loss, (Fig. 5 and Fig. 6) showing difference of contour of audiograms.

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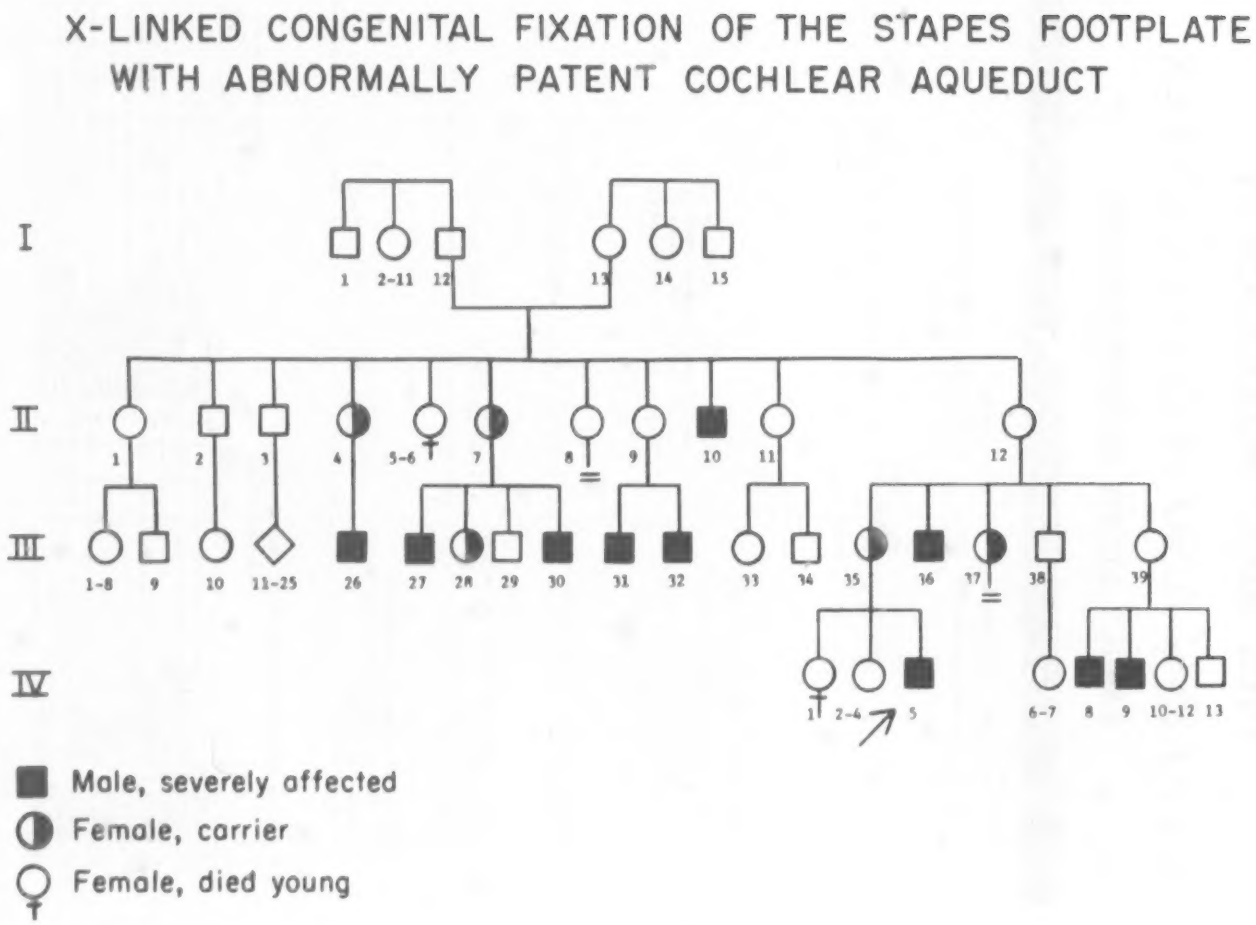
X-LINKED CONGENITAL FIXATION OF THE STAPES FOOTPLATE
WITH ABNORMALLY PATENT COCHLEAR AQUEDUCT
I
II
III
IV
1 2-11 12
13 14 15
1 2 3 4 5-6 7 8 9 10 11 12
1-8 9 10 11-25 26 27 28 29 30 31 32 33 34 35 36 37 38 39
1 2-4 5
6-7 8 9 10-12 13
Male, severely affected
Female, carrier
Female, died young

Fig. 8 Kindred with X-linked congenital fixation of the stapes footplate showing 10 examined affected males in three generations and four examined female carriers.

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THE BILL WILKERSON
HEARING AND SPEECH CENTER

FAMILY STUDY QUESTIONNAIRE

1

1. Which of the following best describes the patient's hearing loss:
   ☐ Hearing loss present since birth
   ☐ Hearing loss developed after birth; if so, at what age?_____

2. Do you know what caused the hearing loss (illness, accident, loud noise, etc.)?_____

3. What hospital was the patient born in?_____

4. When the patient was a newborn baby:

a) Did he have the yellow jaundice?	Yes ☐	No ☐	Don't know ☐
b) Was he an "Rh baby"?	Yes ☐	No ☐	Don't know ☐
c) Did he require blood transfusions?	Yes ☐	No ☐	Don't know ☐
d) Was he premature?	Yes ☐	No ☐	Don't know ☐
e) What was his birth weight?_____			

5. Did the mother have German measles during the pregnancy? Yes ☐ No ☐ Don't know ☐

6. Were the patient's parents kin, for example were they cousins? Yes ☐ No ☐ Don't know ☐
   If yes, what was the relationship?_____

7. Has the patient ever been treated with streptomycin, neomycin, kanamycin, quinine, or large doses of aspirin? Yes ☐ No ☐ Don't know ☐
   If yes, which drug?_____

8. Has the patient ever had any of the following conditions?

CONDITION	CHECK APPROPRIATE BOX: YES	NO	DON'T KNOW	CONDITION	CHECK APPROPRIATE BOX: YES	NO	DON'T KNOW
a) Fainting spells				k) Poor vision			
b) Broken skull or brain surgery				l) Different colored eyes			
c) Ringing or buzzing in the ears				m) Stiff fingers or toes			
d) A feeling of spinning around				n) Goiter or swelling in neck			
e) More than two fractured bones				o) Extra fingers or toes			
f) Unusual skin coloring				p) Kidney infection			
g) Lumps under the skin				q) Pain on urination			
h) White patch in the hair				r) Discolored urine			
i) Clubfoot				s) Blood in the urine			
j) Cleft palate or harelip				t) Heart murmur or defect			

Fig. 9 Page one of Family Study Questionnaire for medical history data.

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2
3
4

Fig. 10 Pages two, three and four of Family Study Questionnaire for pedigree data.

Fig. 11 Photograph of index case in Fig. 8 (congenital fixation of the stapes footplate) with carrier mother. By permission of parents.

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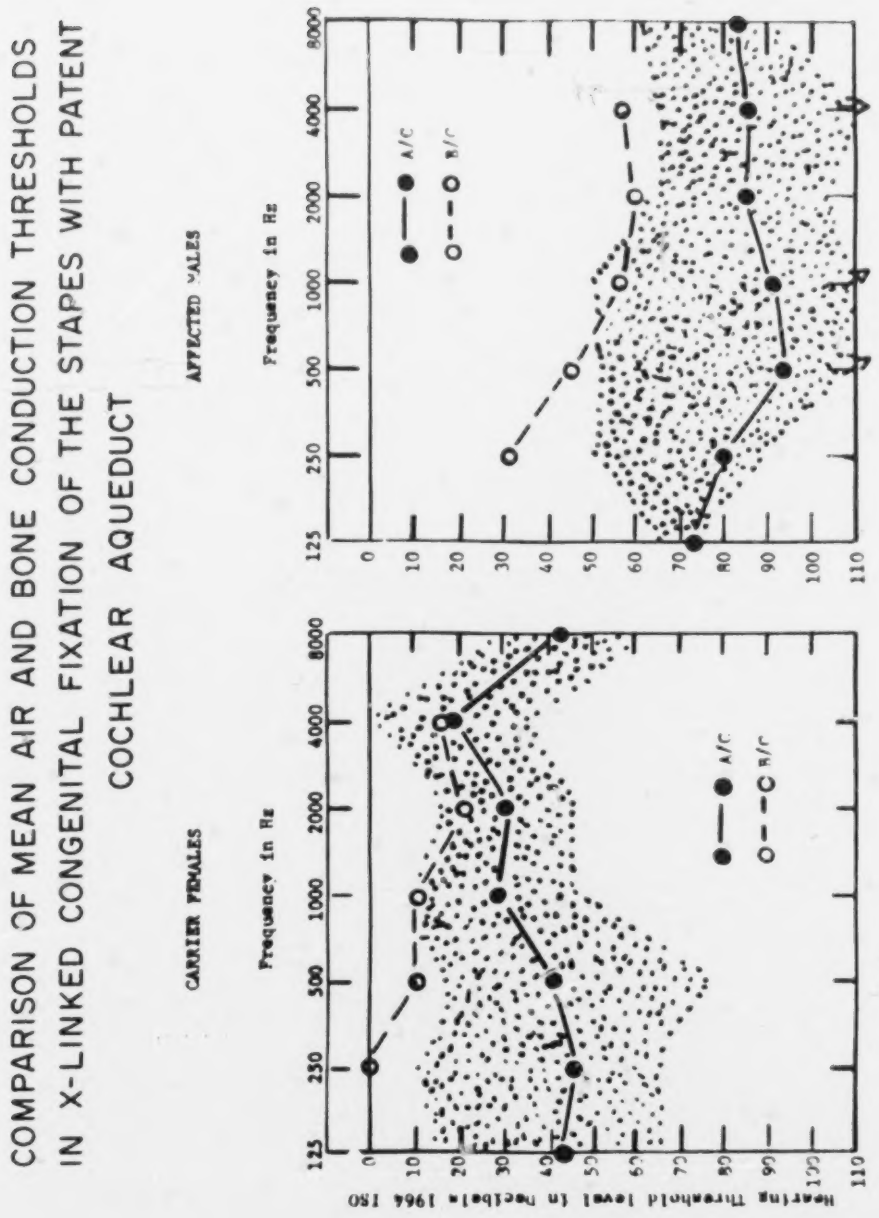


Fig. 12 Composite audiograms of carrier females in Fig. 8 (congenital fixation of the stapes footplate) showing mild expression of the trait in females as compared with composite audiograms of affected males in same kindred.

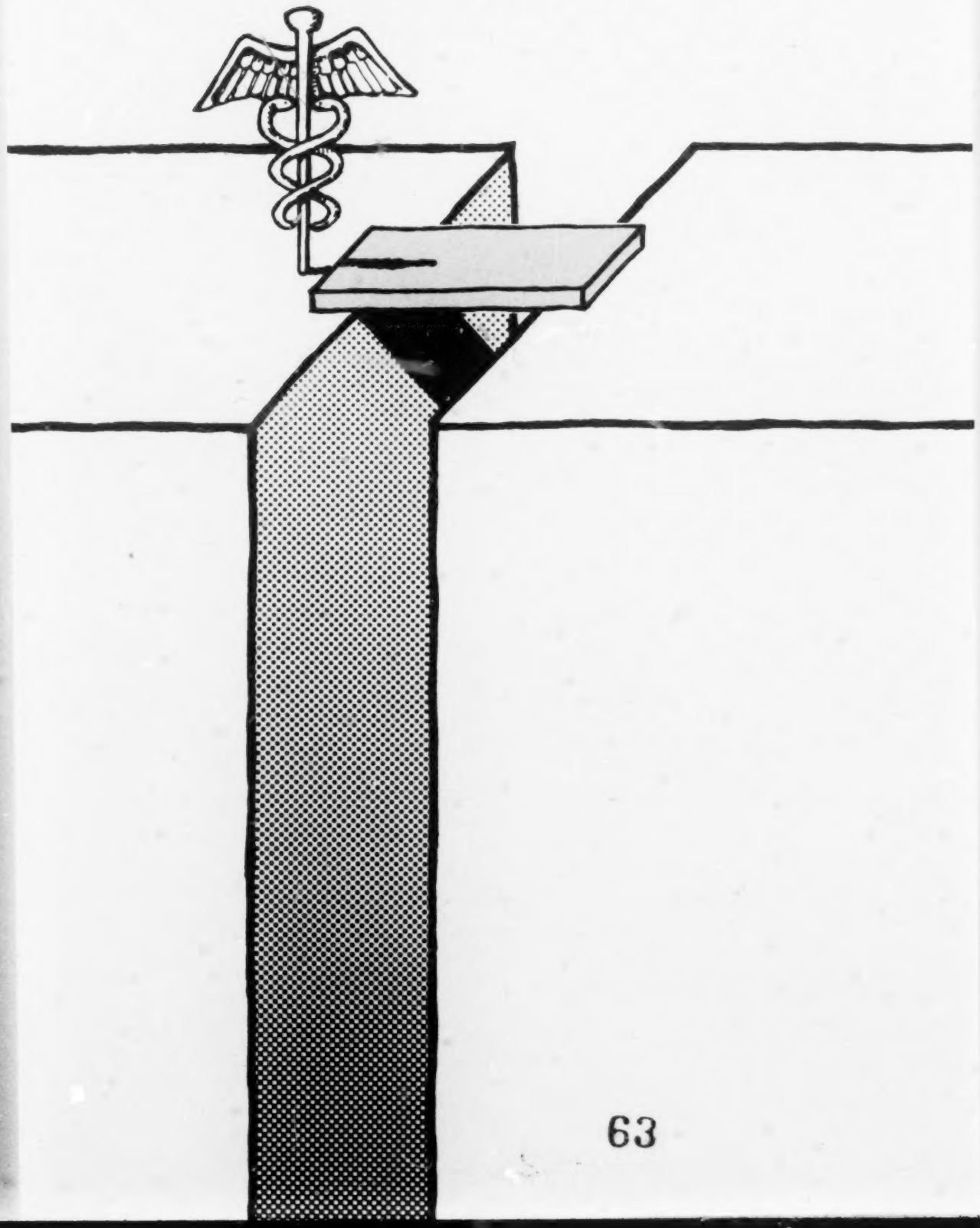
DISCUSSION SUMMARY: PREVENTION

The following concerns relative to the prevention of deafness were identified by discussants:

1. Marriage counseling and genetic counseling are needed for deaf persons and parents of deaf persons. Additionally, training programs in the area of genetic counseling for professional workers with the deaf and for physicians are needed.
2. There is a need for schools for the deaf to offer comprehensive senior high school courses in the area of life adjustment.
3. There is a need for manual communication training for physicians, nurses, and other medical personnal as well as for interpreters to be placed at their disposal when needed.
4. There are geographic areas where additional services are particularly needed, mainly rural and the urban ghettos.
5. There is a need for medical educators to include more information regarding deafness in medical school curricula. Suggested ways to accomplish this include utilizing existing community resources, particularly schools for the deaf, through such avenues as video tapes, demonstrations, and lectures by professionals in the field of the deaf.
6. Educational programs relative to prevention are needed for parents who already have deaf children.
7. There is a need for more and better ways of distributing information, particularly literature to physicians and medical schools. A variety of media might be utilized: audio tapes, video tapes, pamphlets, books, films, letters, and direct contact.
8. Organizations, especially the COSD, need to assume a leadership role in developing and implementing educational programs aimed at Prevention.
9. There is a need for the physician's role to go increasingly beyond treatment of pathology or prescription of prosthetics and to include additional services to the deaf person and his family in the psycho-social areas.
10. There is a need for the development of political machinery to create awareness among medical professionals as well as awareness among appropriate persons on the Washington scene relative to the price society pays for ignoring the needs of deaf persons.
11. There is a need to encourage medical practitioners to be more accepting of deaf clients and to refer them to another agency only when there is special need to do so.
12. There is a need to include deaf persons within the professions of medicine and related health care.
13. There is need for organizations serving the deaf to become increasingly involved in helping solve the problems related to noise pollution.

14. There is a need for an accurate and comprehensive census of the deaf to be maintained on a continuing basis.
15. There is a need for additional research funds to be directed toward projects aimed at the prevention of deafness.
16. There is a need for additional traineeships for preparing professionals to work with the deaf in the educational, psychological, and medical areas.

Diagnosis



CARL W. FULLER, Ph.D.
Assistant Director
Indiana University Medical Center
Indianapolis, Indiana

THE AUDIOLOGICAL DIAGNOSIS OF DEAFNESS

The primary and distinctive role of the audiologist in the diagnosis of deafness is the measurement of hearing. The validity of audiometry with adults is rather easily established in most cases. A great variety of tests is available; so many, in fact, that it might be considered inhumane to subject any one person to all of them. These include tests of hearing thresholds for pure tones and for speech, by air and by bone conduction, with and without masking; tests of speech discrimination; tests of recruitment; of perstimulatory adaptation; of hearing for speech in the presence of a competing message. There are tests administered by live voice and others administered by recorded voice. There are tests to differentiate cochlear from auditory nerve from central nervous system lesions. There are tests to detect the presence of so-called nonorganic deafness, a paradoxical term which means that the patient does not have a hearing loss.

This battery of test procedures has not developed willy-nilly. Rather, it represents recognition of the fact that the measurement of *hearing thresholds* simply does not define by itself the way the auditory mechanism functions. This is a point to which I wish to return, so I will emphasize it now. *The functional utility of a person's hearing can be adequately defined only by a comprehensive battery of audiological tests, not by threshold audiometry alone*. Even the present test armamentarium does not tell us all we would like to know, and so the search for new ways of assessing auditory functions continues.

The test techniques used with adults are not easily applied to audiometry with children. The child's natural anxiety, his short attention span, his limitations of verbal language, his rapid fatigue—these and other factors hinder the testing of children. Nevertheless, audiologists have been successful in devising audiometric procedures and motivational devices which enable very young children to cooperate in threshold hearing tests. Air and bone conduction pure tone thresholds can be measured with relative ease in children down to the age of three years, and in many two year olds. An approximation of a speech hearing threshold can be made even with non-talking children. Speech discrimination scores are less reliable in children, particularly those with little speech. The remainder of the audiological test battery as it is presently used with adults demands an intensity of concentration and a maturity of judgment which even school-aged children find difficult and preschool children cannot begin to understand.

Valid Technique Promised

It now appears that lack of cooperation may no longer be an obstacle to threshold audiometry in youngsters (or in adults, for that matter). The recent emergence of electroencephalographic audiometry (ERA) from the research laboratory and the standardization of procedures for its clinical application promises to provide audiologists with a valid technique for measurement of hearing in children who are untestable by traditional procedures. The particular value of ERA lies in the fact that the acoustic response can be detected in cortical electrical activity even when the test subject is asleep or sedated. It is not a foolproof test technique. Like other audiometric procedures, it requires a skilled clinician for competent administration and interpretation. It also requires expensive instrumentation and, consequently, it is likely to be available—at least in the immediate future—only in centers which have generous funding. Nevertheless, it may well overcome what has been a serious obstacle to confident audiologic practice.

Until ERA becomes routinely available to all children—and perhaps even after it is available—the validity of audiometric findings in children must be confirmed by analysis of the child's behavioral symptomatology. It is well known that some children are indifferent to acoustic stimulation when, in fact, they have normal hearing. The characteristic constellation of behaviors exhibited by deaf children differs in clinically definable and diagnostically significant ways from the behavior exhibited by other children who are not deaf but who are unresponsive to sound. In many deaf children this behavior is so striking that a diagnosis of deafness can be made on this basis alone; the audiometry thereafter provides a quantitative estimate of the degree of hearing loss. Threshold audiometry alone, without supporting behavioral evidence, is rarely sufficient to establish a diagnosis of hearing impairment in young children.

You will note that audiologic testing of children does not achieve the comprehensive exploration of auditory functioning that can be accomplished routinely with adults. The younger the child and the more severe his language handicap, the more likely it is that the audiological assessment will be confined to the measurement of pure tone hearing thresholds, although, as noted previously, speech testing may be possible in some cases. Thus, the audiological assessment of children does not sample other dimensions of auditory functioning which affect the efficiency with which he can utilize his hearing. In children, therefore, the audiologic diagnosis of deafness rests upon only a partial definition of total auditory function.

Different Impact Occurs

We know that deafness has a very different impact when it occurs in a person who has mature language skills than it has upon the person who is deaf at birth or incurs his hearing impairment early in childhood. The differences are familiar to all of you, and I will not dwell upon them here,

except to emphasize that they impose a peculiar burden on the audiologist and his professional colleagues. To explain that statement I need to remind you of what deafness is.

The Conference of Executives of American Schools for the Deaf has defined deafness as a condition in which "hearing is nonfunctional for the ordinary purposes of life." Davis in a more recent definition has suggested what he calls a "social criterion" (as opposed to an audiologic one) "namely, that everyday auditory communication is impossible or very nearly so." The two definitions can be neatly combined: deafness is a condition in which hearing is nonfunctional for everyday auditory communication.

Now let us suppose that an audiologist has completed his testing and that he has accumulated some set of audiologic data which clearly indicate the presence of an organic hearing impairment. Let us also suppose that all conditions which might mimic deafness–psychosis, mental defect, aphasia, environmental deprivation, malingering, etc.–have been ruled out. Then the diagnosis of deafness in an adult is a diagnosis of an essentially static condition. The validity of the diagnosis is virtually identical to the validity of the audiologic findings. We need only ask: "Do these test results show that the person's hearing is functional for everyday auditory communication?" In the case of the adult, if the answer is yes, the person is hard of hearing. If the answer is no, he is deaf.

The same question is pertinent in two situations when dealing with children, but in a third situation the question must be rephrased. When a mild to moderate hearing loss is present in a child, the residual hearing is almost certain to be functional for everyday auditory communication, and the child will be diagnosed as hard of hearing. When the hearing loss is profound, the residual hearing is almost certain to be nonfunctional "or very nearly so" for everyday auditory communication, and the child will be diagnosed as deaf.

In the intermediate range of hearing levels, however the condition presented by the child is not static. In fact, it is most unlikely that the way the child uses his hearing when he is first seen will remain unchanged. A basic assumption of audiology and of education in dealing with children having hearing levels in the range of 60 to 90 dB is that, no matter how the child uses his hearing when he is first seen, he can be taught to do better. Some individuals with losses at the severe end of this range seem to get tremendous amounts of information out of their limited auditory input. Other individuals with losses in the low end of this range, that is, with audiometrically moderate hearing deficiencies, seem to have extraordinary difficulty in acquiring or utilizing vocal-verbal communication even after extended teaching and use of amplification.

Diagnosis of Deafness

A diagnosis of deafness–or of "hard of hearingness"–in this group therefore requires the audiologist to change the tense of his verb. Instead

of asking "*IS* this child's hearing functional . . .?" he has to put his question in the future tense: "Will this child's hearing *become* functional for everyday auditory communication?" If the audiologist feels that this question can be answered yes, he will classify the child as hard of hearing. If he feels that he must answer no to the question, he will classify the child as deaf. Thus, the audiologic assessment of children must be valid not only in terms of the measurement of present status, but it must also have validity as a prediction of future communication skills.

How can the audiologic appraisal of children improve its predictive validity?

The answer, I believe, lies in an attempt to achieve a more comprehensive audiological evaluation of the child. I now return to the point I emphasized early in this paper. I said then, and I repeat now, that the functional utility of a person's hearing can be adequately defined only by a comprehensive battery of audiological tests, not by threshold audiometry alone. And yet too often, with children at least, we persist in putting all the eggs of definition into the single basket of hearing sensitivity. There are other kinds of auditory impairments which affect the individual's ability to use his hearing "for ordinary everyday communication." Three of these–speech discrimination, recruitment, and intolerance for loud sound–can be defined audiologically with relative ease, at least in older persons. All of these disabilities affect the efficiency of hearing aid use. All of them influence ease and accuracy of speech perception. In addition, other factors, such as auditory fatigue, probably play a role in limiting the use of residual hearing for ordinary everyday communication.

I will cite a single example. We once saw a child in the clinic where I work who had hearing levels of 70 dB in one ear and 80 dB in the other. One ear had a speech discrimination score of 70 percent for PBK words; the other ear had a score of 20 percent for PBK words. On the basis of thresholds alone this girl was hard of hearing in both ears, but she had such difficulty with speech discrimination in one ear that she could not wear a hearing aid on that side. Using a hearing aid in the ear with good discrimination, she responded well to teaching, learned to talk, and eventually made successful progress through a school for hearing children. In terms of overall daily functioning she was a hard of hearing child. Yet I submit to you that she was hard of hearing in only one ear; and that the ear with the poorer discrimination could only be categorized as deaf.

It is a well known statistical rule that a battery of tests gives more reliable and more valid results than any single test in the battery. This is certainly true of clinical audiology. The tremendous variety of audiological tests now available permits a very detailed analysis of the ways in which auditory functions are altered in a hearing-impaired person. It should be possible to devise ways in which this battery can be applied to the audiological assessment of children. If and when this is done, the audiological diagnosis of deafness will be more realistically related to the operational definitions of deafness with which we now are forced to operate.

3367

MRS. DONNA PFETZING
Professional Interpreter
Mother of Deaf Daughter
Buena Park, California

DIAGNOSIS: FACT OR FALLACY

Hand clapping, bell ringing, stamping on the floor, and turning up the volume on the radio are all excellent ways to detect deafness in the young child. I should interject here that this is in direct contrast to Dr. Carl Fuller, of this panel. I am probably the country's leading authority on how *not* to diagnose deafness. Yet I find it shocking to learn how many professionally trained medical persons would resort also to the aforementioned methods when asked, "Doctor, does my child have a hearing loss?"

Four years ago, during one of my daughter's routine check-ups, I asked her doctor that question. When he had completed examining her, paying particular attention to her ears, he sat her carefully upon the table top and clapped his hands behind her back. Her curly blonde head immediately jerked around. Satisfied that her hearing was "normal," the doctor laughed at me. He was oblivious to the fact that if she were hard of hearing, she would have heard the clap; if she were severely deaf she would have received some kind of sound stimulus; and if she were profoundly deaf, she would have undoubtedly watched the movements of his arms. So regardless of the degree of her hearing loss, her reactions were identical to that of a hearing child. But I happily accepted the doctor's assurance that nothing was wrong.

Within the next few months, however, the fact that she still was not talking bothered me so I consulted a second physician. Being more sophisticated than the first doctor, he hid a shiny silver bell in the palm of his hand and rang it behind her while she was busily sticking her fingers inside the torn lining of a fuzzy teddy bear. She reacted partially to his dinging and donging, but she was more impressed with the white flakes which were by now quickly pouring out of the side of her furry friend. The doctor told me in a grave tone that her hearing was definitely "low," but that she was much too young to be able to achieve a proper diagnosis. He suggested waiting a couple of years until a more accurate hearing test could be given and evaluated.

I returned home, but an attempt to ignore the situation was useless and before long we were sitting in the waiting room of a third doctor's office. Now, this pediatrician did not agree that her hearing was "normal," nor that it was "low." He simply told me he didn't know, and gave me the number of an audiologist who had experience working with young children, saying I probably should take my daughter there if I was going to continue to worry. Needless to say, I called the number and requested an

appointment, but the waiting list was long and the first available consultation was three months in the coming.

It was during these three months that I became the best diagnostician on my block. The ingenuity of motherhood was ceaseless, and I went about inventing various methods for infant testing that I am sure would set the medical profession back a hundred years. Slamming doors until the hinges bent, blowing whistles that made our dogs howl, and honking the horn on the family car until my neighbors complained, were just a few of the better ones. I would wait until Bobi was quietly playing outside and then sneak up behind her and yell "dinner's ready" or "Daddy's home." I became a familiar figure she expected to see lurking behind every tree and shrub in the backyard–not to mention the number of unsuspecting playmates I scared away!

No Consistent Response

Even when drying the dishes, it was impossible to resist the urge to bang the pans together to see what her reaction would be. The only consistent response I got was from my husband, who was sure by now that I was the one with the problem.

But for all my efforts I was more confused than I had been at the onset of this quest. When she responded to the noises I made, I was sure she heard them. When she didn't I rationalized that she was absorbed in play and simply ignoring bothersome distractions. It was terribly frustrating not to know if she turned around because she saw me or because she heard me.

Then I hit upon the perfect solution, the one foolproof test that would indicate once and for all if she had a hearing problem. I waited nervously for evening to come. Taking a big, loud alarm clock down from the top shelf, I dusted it off and wound it tight. Being sure Bobi was fast asleep, I slowly tiptoed into her bedroom and, standing beside her crib, deliberately pulled out the knob which set the clanging alarm into action: one second, two seconds, three, four, five. No longer able to stand the ringing in my ears, I shut it off as my eyes flooded with tears; her head still lay undisturbed upon the pillow. My realization that she was not only hard of hearing, but that she must be deaf, threw me into a completely hysterical state.

After several hours had passed, my distraught and throbbing head again began to function. But I simply did not want to accept the fact that I might have a handicapped child. My only alternative was to discredit the validity of my foolproof test. This was easily accomplished by setting off the same alarm clock in my son's room. He also slept undisturbed through the noise. This night was the limit of my endurance and I gave up, relinquishing the task of diagnosing deafness to those who were wiser and more qualified and less emotionally involved. However, one month later, the audiologist was to confirm that nightmare. Our daughter was severely deaf as a result of damage to her auditory nerve, which was probably caused by a rubella virus during my pregnancy.

Parents Learn Techniques

Medically, the loss of a few months or even a few years delay in discovering a child's hearing problem may not be considered detrimental. But, educationally, those years between one and six are the most critical for learning language. And since the normal input for receptive language has been blocked, the parents must learn new techniques and skills in order to bypass the auditory system–all of which takes valuable time.

Soon, teaching words becomes the driving force behind every planned activity. Vocabulary building, lipreading, receptive and expressive language lessons, speech, signs, and fingerspelling become a way of life designed to try to fill the void of that silent environment.

I can still remember one time when we took Bobi to the park. Our new word for that day was the expression "hot dog." The entire family ate hot dogs for lunch regardless of whether they wanted them, and we were careful to use this expression at every opportunity.

"Do you like your hot dog?" "I like mustard on my hot dog." "Daddy's hot dog is all gone." I was also concerned with teaching her the correct concept of these words, so I carefully explained that she was not really eating a dog–that this was just a silly name for a kind of meat. I was pleased with the afternoon's events and especially proud of the thoroughness with which we had covered the subject. That evening when she stood in front of the refrigerator door, even though I knew what she wanted, I waited to hear her use the appropriate words, "hot dog." When she looked up at me to see why the door was not opening as she had expected, I asked, "What do you want?" With all the confidence of a child who has learned her lesson well she responded happily, "I want cold dog."

Then there is the story about a good friend who had worked for weeks teaching her young daughter the word for "bathroom." She had made a point not to limit the meaning of the word to just the bathroom in her own home. Consequently they had visited the bathrooms in all the local restaurants, gasoline stations, and even the one situated atop the International Airport. Finally, she was satisfied that the child understood completely the use of that word.

One evening when Daddy was babysitting, he told his daughter to go put her dirty clothes in the bathroom. She gave him a rather funny look and ignored his gesture toward the door. The father, assured that she had understood his command, was determined to control the situation and keep the limits of his discipline rigid. Again he stated in no uncertain terms, "Take those clothes to the bathroom!" The child, seeing that he meant business, reluctantly picked up her small bundle and sullenly trudged down the hallway. The father's face began to beam with the pride a parent feels in his obedient child, until he heard the all-too-familiar sound of the toilet flushing her clothes down the drain. The child had not been mischievous. She had placed her clothes into what for her was the bathroom.

Obviously, the acquisition of language is by far the deaf child's greatest stumbling block. Because he is not bombarded with sounds all of his waking hours, his language development may be extremely retarded. By six years of age a hearing child has almost complete knowledge of syntax and sentence structure. The deaf child at six is lucky if he can combine four words to make a simple, grammatically correct sentence. To illustrate the seriousness of this problem, I would like to read an excerpt from a typical paper written in a Junior High School English class. The author is in the eighth grade, she has an I.Q. of more than 100 on non-verbal tests; she is 16 years old.

WEEKEND

Judy and I went to mountains with my family last Saturday. Judy and I walked look river. She and me say motorcycles. She and me saw car ride down hill. My brother and my friend. John sat on sand. She and me scared of car. My family ate lunch. She and me climbed the mountains.

Last Saturday Judy and me went to bowl with my mother. Judy and I buy fire french and coke. I don't played a bowling because I don't know how to play.

The humor in the four-year-old's mistake is conspicuously lacking when the child becomes 16 and finds reading books on a fourth-grade level difficult, if not impossible, to comprehend.

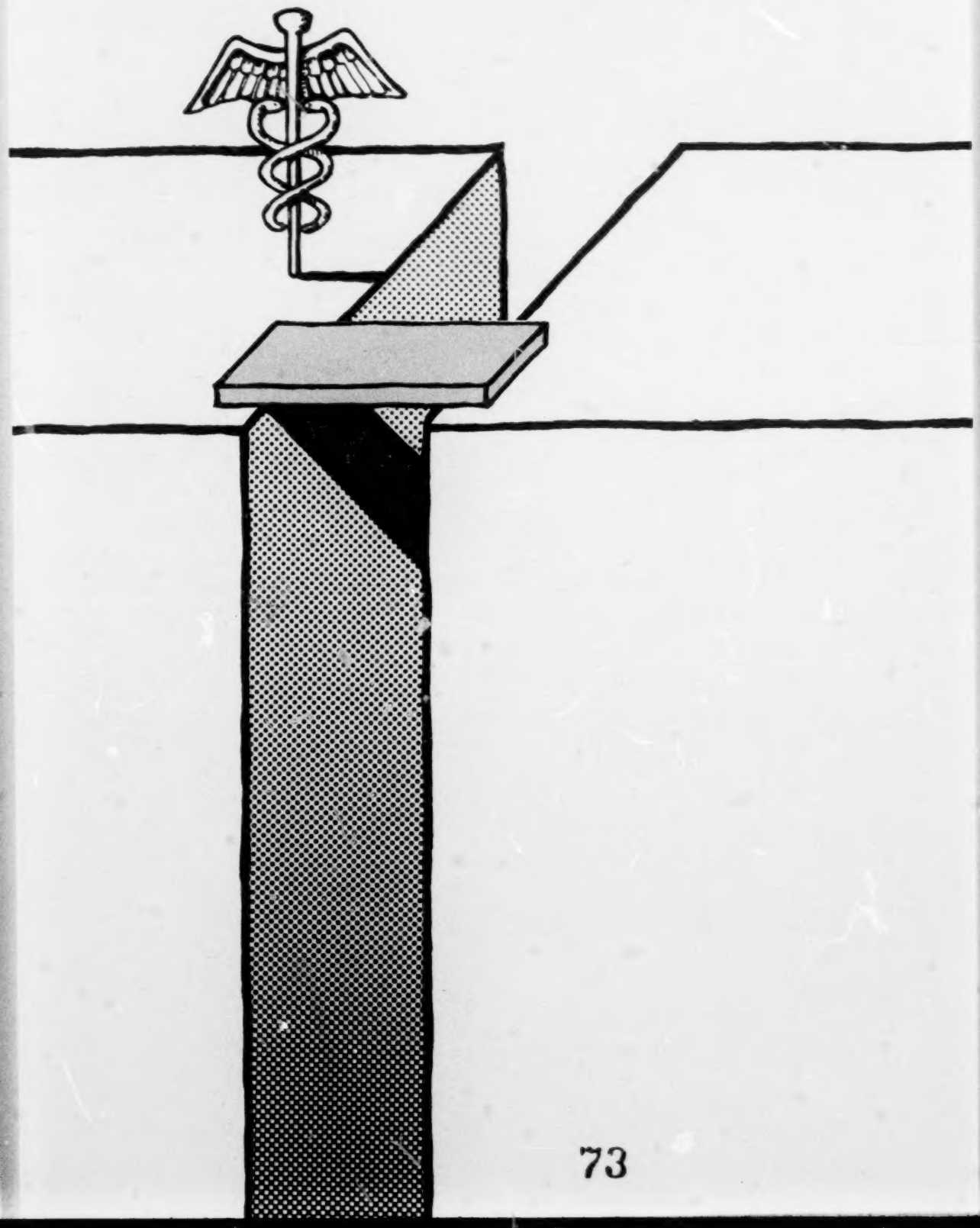
I hope by now the need for early diagnosis of deafness is self-evident. It is imperative if our children are to receive full benefit from early education and communication in the home. They need not feel isolated and rejected if they are being accepted and included in the normal social activities of a busy household, for deaf children are wonders to behold. They are not sad little underlings sitting ignored in some out-of-the-way corner. They are happy, noisy, energetic youngsters who delight in the knowledge of the world about them. They may have to work a little harder to understand your conversation, but there is no communication problem when it comes to a radiant smile minus two front teeth, or a cold nose pressed against your cheek to show you how weather conditions are outside, or a playful bear hug strategically placed to delay going to bed just one minute more, or a soft and drowsy little kiss when awakened from blissful sleep. They are a story just begun of love and laughter, heartbreak and fun.

DISCUSSION SUMMARY: DIAGNOSIS

The following concerns relative to diagnosis of persons having hearing impairment were identified by the discussants:

1. There is a need for greater emphasis for early identification programs in hospitals and in well baby clinics.
2. While emotional upset cannot be avoided, a need exists for programs to lessen frustration of parents of deaf children during the sometimes lengthy diagnostic procedure and to provide continuing counseling and other assistance on a long-term basis.
3. There is a need to utilize more trained deaf persons to assume leadership roles in the dissemination of information regarding deafness, both to physicians and to other professionals, as well as to assist parents of young deaf children.
4. Comprehensive counselling and habilitation services need to be available within the home setting for young deaf children and their parents, particularly during the critical times between the first diagnosis and the time the child enters school.
5. Training programs and other assistance for public health nurses would be helpful in extending services to young deaf children, particularly in large cities and in isolated areas.
6. There is a need for an information center to disseminate information regarding deafness to professional personnel and to provide assistance to parents of young deaf children. Organizations such as the COSD could assist, particularly with the dissemination of literature to medical personnel and to medical journals.
7. Provision is needed for parents of young deaf children to become acquainted with, and knowledgeable about, deaf adults.
8. There is a need for participation by more adult deaf persons on boards of schools for the deaf as well as other boards, advisory committees, and such bodies which are in a position to make decisions about education and other habilitation programs.
9. Parents of deaf children need comprehensive information about all services available to them and to their child and, as well, deserve a share in decision-making regarding all phases of the management of their child's habilitation.
10. There is a need to expand the scope of training programs for audiologists to insure that they have comprehensive and realistic information about deafness and that they are prepared adequately to give advice to deaf persons and to families of deaf persons.
11. There is a need to identify and make known the names of physicians who are interested in having deaf persons among their patients.
12. There is a need to encourage the American Medical Association to establish a specific sub-committee for dealing with the problems of deafness.

Habilitation



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MEDICAL ASPECTS IN HABILITATION

As a medical doctor, who is a psychiatrist, I am indeed honored to be a panelist to discuss the topic of medical aspects of deafness with specific reference to habilitation. To be sure, hearing impairment which affects some 15,000,000 Americans represents the greatest single handicapping condition in the nation. Among these hearing-impaired persons, about 300,000 to 400,000 are considered to be deaf.

Since deafness is a biomedical condition, it is altogether fitting that we should consider this condition from the medical aspects. However, I would like to extend the meaning of medical to include comprehensive health considerations: physical, mental and social. Health is not only the absence of illness or disease but is a dynamic process of well-being. It includes man's interaction with his environment in a way that is satisfying and happy. When we consider habilitation we must first define the term. According to Webster's Dictionary, habilitation is the process of habilitating. The word habilitate means to equip for working or to qualify oneself. Using that definition as a frame of reference, habilitation in deafness from the health standpoint may be considered in five different areas, viz., (1) preventing deafness (2) making hearing possible (3) keeping deaf persons healthy (4) training hearing persons to provide health care for the deaf, and (5) training deaf persons to work in the health field.

Prevention is indirectly a part of habilitation. Knowing the causes of deafness will put us in a better position to take preventive measures. Thus, for those hereditary factors, genetic counseling is coming of age. The rubella factor in the pregnant woman during her first trimester is now being approached through a program of rubella vaccination. Much attention is being paid to the noise pollution factors and steps have been taken to reduce or eliminate these. Many public health measures have been instituted to deal with infectious disease prevention, including those diseases which cause deafness—particularly scarlet fever, meningitis, and others. Toxic conditions and the Rh factor also have been given due consideration.

The second area, i.e., making hearing possible, is centered on surgical procedures (the procedure of stapedectomy used in otosclerosis is quite effective in many cases). Of course, detection of deafness in early infancy is another breakthrough in the area of habilitation. This enables specialists to take corrective action before irreparable harm is done.

Notwithstanding the fact that an all-out effort should be made to prevent deafness or to make it possible for deaf persons to hear, we need to deal with a third issue. We need to make certain that deaf persons receive quality health care services. No one would deny there is a great need to improve the health care delivery system in general. This is more apparent when it comes to services to deaf persons. The number of physicians who would invest the amount of time to deal with deaf patients in face of the communication problem is limited. Even at that, the physician needs to be sure that he is understanding the patient and that the patient is understanding him. This is even more important where the physician is a psychiatrist treating the deaf patient for a mental health problem. Steps have been taken to improve communication. Accordingly, the Registry of Interpreters for the Deaf has prepared informational pamphlets for physicians and specialists in the field of mental health. This represents a valuable contribution in acquainting the health care worker and the interpreter with the communication problem and a way of dealing with it. However, the use of an interpreter is not completely satisfactory, because it makes a third party necessary in a situation where privacy is so desirable.

More Knowledge Needed

This, then, brings us to the fourth consideration. Problems could be dealt with more effectively if health care workers who have normal hearing—including physicians—were more knowledgeable about problems of the deaf and who possessed special skill needed in communicating with them. The value of such an approach was made very clear to me from one of my own experiences in working with deaf persons. The example refers to a deaf couple who had three hearing children ranging in ages from six to 12. The middle boy, age nine, was experiencing a life-threatening weight loss due to his refusal to eat. In addition, he was experiencing learning difficulties in school. The pediatrician worked with the boy and his family trying to correct the problem but was making slow progress. He realized the communication difficulties in dealing with the patients, but he also realized that it was necessary to work with the parents in order to effect positive change in the boy. The pediatrician also recognized the emotional and mental health factors involved with the family and referred them to a Mental Health Studies Center operated by the National Institute of Mental Health. The child psychiatrist there immediately determined that the family could best be served by mental health workers who had experience and communication skills in dealing with deaf persons. Therefore, he referred the family to the Mental Health Program for the Deaf at Saint Elizabeths Hospital. Working with the family from appropriate frames of reference were, in addition to the pediatrician, a psychiatrist and a social worker, both of whom were skilled in communication with deaf persons and knowledgeable about mental health problems of deafness. The effect of this team approach was the rapid resolution of the immediate problem.

The boy improved in his eating, he gained weight, and his school work improved, also. Beyond this, interpersonal relationships within the entire family improved.

In improving health care delivery systems, more health care manpower is needed. It is estimated that by 1980 we will be 25 percent short of the 1,776,000 needed health care workers who require college training. Of those not requiring a college education, we will need 580,000 by 1980 but only 475,000 will be available. How many of the total needed will be skilled in working with deaf persons? Obviously, there will be few. Therefore, more education and training must be directed toward preparing and equipping health care workers to deal specifically with problems of deaf persons. Because congenital or early acquired deafness imposes a particular life style on an individual so affected and tends to isolate him from the main stream of society, particular attention must be paid to the health needs of this particular group. Health care workers must become more aware of the implications of deafness, such as communication problems, social isolation, problems of vocational adjustment, special educational procedures required, prejudices against deaf persons, etc., and how all of these factors bear upon the total health needs and delivery systems available to them. To be effective, the health care worker must be indoctrinated in special methods of delivering health care to this group.

Health Care Occupations

In the United States there are about 125 different health occupations. Exactly how many of these include services to deaf persons is not known. Training in health occupations is generally available to hearing persons and even more attention is given to minority groups now than before in helping them obtain such. However, such training for deaf persons still lags. I take some pride in feeling that in some way I have been responsible for training a sizable number of hearing and deaf persons in providing health care, including mental health care for the deaf at Saint Elizabeths Hospital.

Finally, the fifth consideration in habilitation is training deaf persons in the health occupations. Some schools of higher learning do provide such training, on a limited basis. However, it is exceedingly difficult to find that a deaf student has ever been accepted to medical school for education leading to the M.D. Degree.

Is it possible for a deaf person to become a medical doctor? This question was asked of me by Mrs. Edna Adler of the Department of Health, Education, and Welfare in April, 1967. My affirmative answer at that time has since been markedly reaffirmed because of my experience in teaching and training health care workers, both deaf and hearing. This experience has covered a span of 15 years and has included teaching in medical schools, a school of social work, and in the Department of Psychology at Gallaudet College. Moreover, I have supervised the training of deaf and hearing professionals in the conduct of psychotherapy in the Mental

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Based on my experience during the past seven years in teaching and training, particularly of deaf persons, I feel there are deaf persons who have the potential for medical training and, if given an opportunity and all of the supports, could succeed in becoming medical doctors. Such supports may include, but would not necessarily be limited to, visual aids (slides, films and video tapes with captions or overhead transparencies), electronic devices with visual indicators to substitute for listening, interpretive services, hearing study mates, tape recordings with hearing persons available to transcribe or interpret later, counselling, and financial aid. Many medical schools are now modifying their curricular to provide more appropriate training so that the student will be prepared to give better service.

It is estimated that about 50,000 more doctors will be needed by 1980 to provide adequate health care. I feel, therefore, that with the recent impetus in modifying medical school curricular to increase medical manpower and to make training more appropriate to meeting service needs, the time is ripe for deaf persons to capitalize on this. Many medical schools work in conjunction with colleges and universities so that students can complete portions of the curriculum requirements in these colleges and universities and enter the medical school program with advanced standing. I have proposed this idea to Gallaudet College and to two medical schools in Washington, D.C., and the proposal has received favorable consideration.

If efforts such as these can succeed, I feel that we will have made a valuable breakthrough in the health care field which would have far reaching implications in comprehensive health care, including habilitation in deafness.

Due recognition needs to be given to research efforts which have contributed to our present body of knowledge and skills in the medical aspects of deafness and the need for continuing research. However, a more detailed discussion of research aspects is not within the scope of this paper.

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A NURSE LOOKS AT THE HEALTH PROBLEMS OF DEAF PEOPLE

It seems to me that an active practitioner of nursing with 30 years of experience ought to be able to speak to you with considerable authority and expertise about nursing care and the health problems of deaf persons. Yet as I approached the composition of this address to you I became more and more aware of the limits of my knowledge. It was interesting to me to realize that during the first 22 years of practice I had never encountered a deaf patient. Although I had my basic education and training in a 3,000-bed hospital in New York City, I cannot recall any specific curriculum content that dealt with the nursing care of the deaf patient. In a city the size of New York there must have been deaf persons who needed care. Who took care of them? What kind of care did they receive? These questions are of even greater importance today. Who does take care of deaf patients? What kind of care are they receiving?

My state of blissful ignorance about the nursing needs of deaf persons, was abruptly shattered in 1963 when I began working in a sociological study of the deaf and hearing in Frederick County, Maryland.[1] Although my purpose was to develop some skills in sociological research, this purpose became secondary when some of the respondents in the study discovered I was a nurse. Frequently I was asked, "What are you doing about the health needs of the deaf?" In order to answer the question I found it necessary to determine first precisely what were the health problems of the deaf. This determination, as you might suspect, was more difficult than it sounded. Preliminary findings in the Frederick study indicated that persons who work with the deaf are exceedingly concerned with the problems of their physical and mental health. Several areas of major concern were expressed. Among these were the following:

1. Lack of education by the medical and paramedical professions, especially in the areas of promotion of health and prevention of disease.
2. The emotional health problems of the deaf, for example: (a) Who prepares the deaf child for the usual surgical procedures such as tonsillectomy and adenoidectomy? (b) Who preapres the deaf child for the trauma associated with repeated evaluations in clinic situations, such as audiometric tests? (c) Who preapres the young married girl for the problems of pregnancy and childbirth?[2]

These preliminary findings indicated the need for a specific study focused on health needs of the deaf, if we were to come up with any answers based on facts. As a result, a small grant application was submitted to The National Institutes of Health. The application was approved. The study, "Meeting the Health Needs of the Totally Deaf" was conducted in Frederick from January 1964 through December 1964.[3] The study proposed to review as systematically as possible the practices and problems encountered in the medical and paramedical professions in dealing with the deaf in the state of Maryland. It also focused upon the means employed by health personnel in solving the problems of communication in interaction with deaf patients and the evaluation of their success in solving the problems. The health attitudes and experiences of some deaf persons in Maryland were also explored.[4]

What kind of care do deaf patients receive? Is there a difference in the perception of this care as viewed by the deaf patient who is the recipient and the hearing nurse who provides the care? There is a difference as you all know. The nursing care given to deaf patients is most often the same kind of care which adequately serves the hearing patient. It is planned and implemented by nurses whose education and experience foster service to hearing patients. If this service does not meet the nursing needs of the deaf patients it is as frustrating to the nurses as it is to the patients. Nurses can and do modify care. Most often they do this automatically as they observe the handicaps of the patients they serve. Deafness is not a visible handicap. It does not elicit the kind of response that blindness, for instance, calls forth. Nursing today, as well as being highly technical, is a highly verbal process. But you do not hear us and we do not know your language. The barrier of communication keeps us both from reaching our goals—you, from receiving satisfying and comforting care; we, from giving care based on your needs.

Some Anecdotes

Those of you who are deaf will readily identify with the patients in the following anecdotes which illustrate the paradox in nursing care which I have just discussed.

1. A middle-aged deaf man was admitted to the hospital for emergency surgery. In addition to deafness he had a problem with sight. He wore bifocals and carefully placed them on the bedside stand along with his pad and pencil. His usual mode of communication with the hearing world was through writing. Following surgery he required intravenous fluids. After a careful examination of both arms to find the best vein, his right arm was immobilized and the needle for intravenous infusion was inserted. As a result he could not reach his glasses and could not see what was going on around him, nor could he communicate by writing. No one could understand why he was so upset and he had no way of telling them.

2. A young deaf married woman after she delivered a live healthy baby was left alone in the delivery room while still under anaesthesia. The nurses were busy but checked her at frequent intervals. When she awoke there was no one there and she had no knowledge of what had happened. She panicked and became hysterical and unmanageable. Since she could neither speak nor hear she could not convey her fear and concern. The staff on the other hand could not understand the reasons for her outburst.

There are many, many more such incidents. Some terminate with restraint and sedation. Rarely do they terminate with understanding and satisfaction on the part of patients and staff. Why is there so little understanding of the deaf person in sickness and in health? What are we doing to change the picture?

A review of the nursing literature shows a paucity of articles relating to the deaf prior to 1960. Since 1960, however, more and more articles about the problems of deafness are being written by nurses and published in the nursing literature.[5,6,7,8,9,10] Graduate students in nursing are becoming more interested in meeting the needs of deaf patients. They are becoming involved in studies which demonstrate how nurses can work effectively with the deaf mentally ill. Two noteworthy examples are the studies done by Halcomb[11] and Carty.[12] More nurses are working on special units for the deaf, and making the effort to learn manual communication as may be seen at the St. Elizabeths Mental Health Program for the Deaf. Nurses are committed to the service of patients. Today, more and more, we are seeing signs of that commitment to the deaf patient. It will continue.

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HABILITATION–MENTAL HEALTH

All too often one becomes enmeshed in a jungle of professional jargon, so let me explain at the beginning that my focus from a habilitation standpoint will be on mental health. I am not speaking in terms of a dysfunction but rather of the need for a developmental state of well-being, of relevant ego-nourishment, and of a healthy self-concept. This of course means a realistic acceptance of and identification with deafness–both for the deaf person and his hearing associates.

Why is there a pressing need for habilitation of the deaf? And what is the cause for that?

Many of us will agree that the need and the cause can be traced to the fact that most deaf persons are ill-educated. In the field of education "ill-educated" has come to be synonymous with inability to read and write.

Let us probe a little deeper. Joanne Greenberg in her book "In this Sign" mentioned that the memory of Abel, the deaf character, was faulty because there were no words upon which to hang the memory. Let me repeat this: "no words upon which to hang the memory." The thought slowly sinks in that it is nearly the same as saying one has no soul.

Let us go even further and mention emotions. Without the checks and balances of sound, what is a deaf person thinking when someone's laughter seems to be directed at him? What is a deaf person's feelings when there is a sea of communication around him and he is not a part of it? Perhaps the following anecdotes will bring home to you the direction pent-up emotions can take:

> A deaf adult employed by a firm involved in computers and peripheral equipment suddenly told a fellow deaf employee "I wish I had a machine gun. Rat-a-tat-tat, I'd kill all these hearies."
>
> A student returned to school highly agitated. She was like a cornered animal, coiled and ready to strike. "My father, my father," was all she could say. A few minutes later her father drove up. He was in tears. Again and again he said "I can't make my daughter understand."

What is done to a deaf person's self-image when there is the constant and subtle pressure not to accept the fact one is deaf but to strive to be like those who can hear? Even "How to Teach the Deaf" textbooks over-emphasize the importance of sound while relegating the development of powers of observation and visual orientation to secondary roles. Teacher trainees, for example, are told that language learning depends on recognition of stress and rhythm and on the utilization of amplifying

equipment. The accent is on what hearing persons assume the deaf hear and not on what is actually heard.

Society Lacks Tolerance

What does a deaf person think of a society which has demonstrated a lack of patience or tolerance for those who are different? Does a smile flicker at the corners of his mouth as he watches the worship at the shrine of cosmetology? Why wear glasses when there are contact lens? Drummed into his consciousness are advertisements proclaiming hearing aids so small they are invisible.

My stream of questions should raise another question: with all I have mentioned why are not more of us deaf persons cutting paper dolls? That is a good question.

We have become so enmeshed in the methods war that it appears we are thinking of a mechanical apparition instead of a person. Instead of habilitation during the developmental stage we have to fall back on a series of rehabilitations after the deaf person leaves school.

Communication is defined as "Sender Encodes Message and Selects Channel" and then "Receiver Decodes Message." There is an assumption—our field is full of assumptions—that receiver gets the message. Actually, it is one-way communication.

Communication is participation.

My 23-month-old deaf girl is a participant in the communication process. She couldn't care less about the methods controversy. She loves candy, ice-cream, and cookies and quickly learned the signs for them. The degree and type of her thirst fluctuates so she learned the signs for water, mike, and orange juice. She learned to associate the sign "beautiful" for a flower and for her new coat. When she became ill she pointed to her ear and used the sign "hurt." When her daddy dropped an ashtray she looked up at him and used the sign for "dumb." At a certain period in time, communication became intensely meaningful to her. It met her needs and was a reference point for the world around her. She had something upon which to hang her memory.

With my own deaf child there has been a minimum of frustration and the bottling up of emotions. She is not just a happy baby but a joyous one. Thus in my home habilitation has been a continuing process. More important, it has begun at an impressionable age and at a critical time. Incidentally, you might be interested to know that I practiced discrimination. I hired a deaf housekeeper.

We need to habilitate and rehabilitate the thinking of the persons in the deaf person's environment.

Realistic Outlook Necessary

We should agree that parents need a more realistic outlook on communication. They need to be given a bagful of communication tools so

that the appropriate one can be selected depending upon the situation and upon the child's ability and level of readiness. The parents need a greater arsenal of information. They need to know of alternative sources of action to take in order to cope with a squirming, ever-changing, emotional bundle of young humanity.

The medical profession needs to be aware that the medical and educational aspects of deafness are two horses of a different color. An uneasy feeling persists in the medical profession itself that the approach to patients has been too antiseptic, too cold and impersonal. Since the medical profession is the first contact parents usually have with "professional" advice, a plan of action should begin here.

Who is going to do it? A study of minority groups indicates that we deaf persons will have to do our own thing. Of course, our hearing friends are more than welcome to pitch in.

Now, the larger question remains: What is the role and responsibility of organizations of and for the deaf? Bogged down in controversies, organizations have been unable to utilize their resources optimally for the habilitation of the deaf. For positive action to take place there must be a bone-chilling sense of reality. It is useless to argue with what was and will be. What counts is what is. Deaf children and deaf adults do use manual communication. Ignorance of this reality or refusal to face or understand it by parents, those in positions of influence and authority, and others, have caused our manpower, finances, and other resources to be utilized in an ineffective and helter-skelter manner.

Until the time comes when the basic problem of communication is recognized, few deaf persons will stand a chance of being habilitated—and the supreme irony of our field will remain the polarization of the deaf themselves and those who are supposed to help them.

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THE DEAF COMMUNITY'S RESPONSIBILITY IN MEDICAL HABILITATION

We have all sensed throughout this *Forum* the bitter frustrations deaf persons experience in seeking medical habilitation and treatment. It has been said that physicians do not take the necessary time to write, that they do not explain, that they should learn to sign, that they should acquire more information about deafness, etc. The physicians who have taken the time to come here to the *Forum* or who, like Dr. Robinson, Dr. Schlesinger, and Dr. Rainer have almost single-handedly established medical (psychiatric) services for deaf persons must feel somewhat like scapegoats or like Daniel in the lions' den.

Rather than persist in criticizing the physician, I would like to look at the other side of the coin and discuss what we in the deaf community and general public should do. One way to begin is with a case history.

About 20 years ago in a hospital in one of the largest cities in the United States, an intelligent, sophisticated deaf mother gave birth to a baby. It was known that there was present the Rh factor in compatibility, but this modern medical facility had the expertise and equipment to treat the problem. The baby was slightly jaundiced at birth, but the deaf couple and their newborn infant were permitted to go home shortly thereafter. The parents, who had no interpreter, brought the husband's mother along for the final interview with the doctor. This woman heard the physician explain that, if the child's coloring became yellow or there was other evidence of jaundice, they should bring the infant for emergency treatment immediately. They were to come back for a blood checkup in a week regardless.

The grandmother felt that she knew all that was necessary about child care and only interpreted part of what the doctor said. In a matter of days the child became seriously jaundiced. The deaf parents, not having been told the full story, did not return to the doctor, but frantically treated the infant with the grandmother's home remedies. When the time for their appointment with the doctor came, they did not go because the grandmother had not told them they should. The doctor tried to call but there was no telephone.

Finally the child became so intensely yellow and frail that the parents overruled the grandmother and took the infant to the hospital. Blood transfusions were given immediately and other treatment instituted. The infant's life was saved, but this child is today deaf, severely mentally retarded, and has to be strapped into a wheelchair to be moved.

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Anybody who has lived with deafness can relate case histories similar to this from their own experience such as those which Dr. Douglas has described.[1] In the case of the jaundiced child, we can blame the grandmother for her stupidity, we can blame the doctor for not learning to sign, but what about ourselves? What could we have done to prevent this tragedy and what can we do to prevent others like it which, unfortunately, occur alarmingly often?

Interpreting Services

Is it not more realistic for us in the deaf community to use our organized influence to have interpreters provided than to expect doctors who see one or two patients a year to learn to sign? For example, what have we done to try to influence medicare to include fees for interpreters? What have we done to induce health insurance plans to include interpreting costs as part of the benefits?

All large hospitals list staff who speak various languages in order that they can be called on as interpreters when there is a patient who speaks a foreign language. What have we done to notify these hospitals that they should also list staff who know sign language? What have we done to provide these hospitals the names of interpreters? What is the role of the Registry of Interpreters and how has this role been met?

Deaf Professionals

Ironically, while many of us have been complaining about medical services it has been a physician, Dr. Robinson[2] who has moved to open medical schools to deaf applicants. A few deaf physicians in major urban areas is no panacea to health care needs, but it would be a major step forward. Deaf psychiatric nurses, also proposed by Dr. Robinson, would be another progressive move.

What have we done to follow up Dr. Robinson's efforts? We need to locate able deaf students for healthcare positions. We need to encourage federal scholarships for these young deaf persons. It can be done.

My own wife, who is deaf, got herself a "scholarship" of sorts by marrying a man who would support her and do some interpreting. On this scholarship she completed a year of medical school as part of her graduate work in microbiology. She has occasionally expressed the view that earning a federal scholarship, had it been available, would have been preferable to the matrimonial "scholarship" that has involved such a long and burdensome repayment schedule.

Summary

In sum, it is clear from this *Forum* that frustration and anger characterize the experience many deaf persons have in seeking medical habilitation. The point to be made is that the deaf person is the one who pays the ultimate price in medical habilitation because it is he who gets the

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inadequate health care. If it is he who pays the price, then the greatest concern, motivation, and payoff for improving care is his. If major progress is to be made, the responsibility is ultimately going to fall in large part upon the shoulders of the deaf community. This *Forum* represents a major initial attempt to bring about progress in medical habilitation in an organized constructive way. The issue now becomes, "What will be the follow up?"

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DISCUSSION SUMMARY: HABILITATION

The discussants focused on the area of mental health and identified the following needs related to deaf persons:

1. There is a need for mental health centers for the deaf. Such centers should be decentralized, at least to a regional basis, but with additional emphasis placed on involvement of and within local communities.
2. There is a need for a directory of psychiatrists and other professionals in the field of mental health who provide services to deaf patients.
3. There is a need for political forces to be brought to bear on the problem of establishing and maintaining mental health services for deaf persons.
4. Additional services are needed which would assist the deaf person to maintain an acceptable state of mental health after recovery from a mental illness.
5. Provision is needed for recruitment and training of deaf professionals to work in mental health and related fields. The para-professional occupations were suggested as an appropriate initial thrust for greater involvement of deaf persons.

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